

REPORT:

Commission for the Control of
**HUNTINGTON'S
DISEASE**
and Its Consequences



Volume IV, Part 4: Public Testimony —
Denver, Los Angeles

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P R O C E E D I N G S

MCKHANN: I'm Dr. Guy McKhann from Johns Hopkins, one of the Commissioners, and we're glad to see you all here. We didn't have to come all the way from the East to enjoy your weather with you, but we're here anyway.

I'd like to start by introducing the other Commissioners who are here. On my immediate right is Marjorie Guthrie, who may be known to many of you; Dr. Li from the University of Pittsburgh; and Dr. Nancy Wexler on the end, who has been not only a member of the Commission, but also, really, the person who does all the day-to-day work in keeping us functioning.

I'd also like to mention that we have a guest-observer in the audience, Dr. Donald Tower, who is the Director of the Neurological Institute of the National Institutes of Health. He has joined us today just to see how our Commission functions and to get an idea or a preview of the kind of recommendations we're going to be making.

Let me say a few words at the start about our Commission activities and what we hope to get out of these public hearings. We've been in existence now about eight months. We expect to finish our work this summer with a report that will go to the Congress, to the Secretary of Health, Education and Welfare, and to President Carter and his representatives.

We meet every month for a couple of days to discuss particular issues which we feel should be included in this report and we also have had public hearings around the country. We've had four or five prior to the first of the year and now are going through a second wave. A list is in this yellow handout of all the places we've been. We've broken the Commission up into small groups so that two or three Commission members go to two or three different areas. Now the reason why we're doing this is to get some feel for the kinds of problems people who are dealing with Huntington's disease have in various parts of the country.

What we want from you today is input to us into the kinds of problems that you're having, either from the research point of view if you're a scientist; from the care point of view if you're a patient or a patient's family member; or from the point of view of facilities if you're a person who is involved in medical care; and maybe others as well. Now we'd like to hear what your problems are. We'd also like to hear what solutions you can suggest because that's really what it's all about.

We've had a fair amount of input now, and I think we have a pretty good idea of the scope of the problem, but we, like you, are searching for answers, and any input you can give us in this area is particularly helpful.

Now, what we will do is, I have a list of people who have asked or requested to testify before us. We have a table over here and I think we'll ask people to come up in groups of three, although they may not be three people who thought they were going to be testifying together. We'd like to ask each person to limit their testimony to five minutes or so, so we could have five minutes with each person to discuss other aspects and questions and answers that come out of that particular testimony. We'd like to ask each witness to identify themselves, their name, their address, and their affiliation with an organization or why they're particularly interested in this problem. This is being recorded and transcripts of these hearings are available to us later and parts of them will be part of the final report. That's why the microphones, so please speak as clearly as you can into them when you use them.

So, I think, with that background, we might go ahead and start. The first three people that I have on my list are Dr. Ferguson, who is from the Governor's Office, Dr. Lubs, and Dr. Chisholm. So maybe the three of you could come on up and we'll go ahead and start. Dr. Ferguson, you might go first, if you would.

TESTIMONY OF
DR. FERGUSON
DIRECTOR, CHRONIC DISEASE CONTROL
STATE DEPARTMENT OF HEALTH
COLORADO

FERGUSON: Good morning. I bring you greetings from the Governor's Office and his regrets that he could not be here today. He is out of town. I bring greetings also from the State Department of Health and Dr. [Tony Robbins], the Executive Director. That's actually where I am, rather than the Governor's Office. I am the Director of Chronic Disease Control there.

My comments will be very brief. We are very much interested in this meeting, delighted to be asked to participate.

Our Chronic Disease Control Section of the State Health Department is very new. We are 21 months old today, exactly. We are primarily, at the present time, involved in the areas of heart disease control and cancer, to just take number one and number two as far as the killers and disablers, both across the country and in the State.

I apologize ahead of time, but I'm not going to be able to spend the entire day with you. We are unfortunately involved in a hassle with the State Legislature over the support of a particular cancer program. So, about the middle of the morning, I'll have to excuse myself and get back to the battle.

That's about all I have to say. I'd be happy to respond to questions or comments from any of you or from any of the panel.

GUTHRIE: I would like to ask, do you have hopes or intentions of doing anything for families with chronic disease other than heart and cancer?

FERGUSON: Very much. I'm very glad you asked that. I have been in Colorado myself a little bit less than two years. I've been out of school about 10 years. Before I came here I was associated with the State Health Department in Oklahoma, and there we built a complex of 63 genetic counselling programs across the state. There was genetic counselling available through all the county health departments in Oklahoma. This started out several years ago with sickle cell screening but it has now continued into a number of other areas. This is an interest of mine and, I think, an integral part of chronic disease control from a state health department standpoint.

MCKHANN: What do you see, Dr. Ferguson, in terms of the limitations of getting that going; just the funding aspects or are there others?

FERGUSON: The limitations that I see right now are the reality of dealing with--and I'll be quite candid with you--with politicians who do not even seem to see the value in heart disease control, which is the number one killer in this State, and cancer, which is the number two killer; primarily, a problem of education of our legislators and other political leaders, and the development of funding and support thereby.

GUTHRIE: I don't know if you would give us the opportunity, but some of us might be very willing to speak to your state legislators and explain to them some of the long-term problems involved with chronic neurological disorders and genetic disorders which last a whole lifetime for so many people, while cancer and heart usually come at the later years of a person's life. Perhaps we ought to do some cost evaluation and give them an opportunity to learn from some of us who know.

FERGUSON: Very good. I'd be delighted to talk about that.

N. WEXLER: We had some conversations with representatives from Senator Kennedy's office about the interest in chronic diseases at the Federal level, and the Commission was somewhat disheartened by this person's response, which is that he did not see any great burgeoning of interest in chronic diseases or chronic neurological diseases in particular. I wonder if you would have any suggestions at your state level as to how we might, both from the consumers' level--people here--and from our Commission, to increase the interest in these diseases?

FERGUSON: This is a very difficult point. I don't know that I could say anything that will be of any help at all. A few years ago I had a unique experience, to tell a very brief story. A young lady stumbled into my office at the University of Oklahoma Medical School and collapsed unconscious on the floor of my office. And it turned out that she had diabetes and she was pregnant; it turns out that her husband was diabetic as well. She was the secretary of a man in the state who was known as perhaps the diabotologist, if there is such a person, in the State of Oklahoma. But no one had ever mentioned to this lady and her husband, who both were holders of--between them--three Masters Degrees--no one had ever mentioned anything about the genetic problem that these two people, having had juvenile-onset diabetes--no one had ever mentioned this to them and the implications of their having children.

We simply--most physicians simply do not think of genetic diseases; we don't think of chronic diseases, really, until the disease is there. And it's a care problem. Being with the Health Department, my interest is preventive medicine. How can we prevent these things?

I wish I could answer your question but I'm afraid I don't know the answer.

N. WEXLER: Are you developing any particular educational programs, then, for physicians and consumers?

FERGUSON: Within the constraints of the resources that we have, yes, both in the area of heart disease--specifically hypertension control because it's such an important factor in the development of heart disease--and in cancer. Those of you who listen to radio stations, you know I've been on the radio for the last two weeks talking about irradiation and thyroid cancer. I know you're tired of it; I am too.

N. WEXLER: One last question. Is care a part of your program; planning care facilities, or inspection of care facilities, development of new facilities?

FERGUSON: Well, certainly, the inspection of the care facilities is one of our legal responsibilities. We are involved in and interested in the provision of good care across the board. This is something that is changing, though. I think our classical role, as VD clinic, TB clinics, immunization clinics, is changing. We are now into the chronic disease control. This is going to require getting involved in the provision of care as well.

MCKHANN: Dr. Ferguson, as you comment on the status of chronic disease facilities or programs in the surrounding states--we're here on a regional basis--you're speaking about Colorado; you've mentioned some aspects of the situation in Oklahoma. What about other states in this particular region?

FERGUSON: I'm really not familiar enough to speak to that.

MCKHANN: Okay, thank you.

GUTHRIE: I just want to say thank you. Since Woody Guthrie comes from Oklahoma, I'm delighted to hear that you're doing very well there in genetics and hope you can do as well here in Colorado.

FERGUSON: Thank you.

MCKHANN: Our next person is Dr. Herbert Lubs, who is an Associate Professor of Pediatrics at the University of Colorado.

TESTIMONY OF
HERBERT LUBS, M.D.
ASSOCIATE PROFESSOR OF PEDIATRICS
UNIVERSITY OF COLORADO

LUBS: Thank you, Dr. McKhann. I was very pleased to have the opportunity to talk with you all. I've been working in the area of medical genetics and inherited diseases for about 20 years now and we deal with perhaps 2,000 different inherited disorders. But I think from my own experience, and probably also from my colleagues, that we all feel that this is one of the highest burden disorders of this group of 2,000 unusual and difficult diseases. So, to have the opportunity to help the cause in some way is very gratifying to me.

What I would like to do is just to summarize very briefly some of the problems that I've run into in working with families, both from the standpoint of the medical school and from the standpoint of the families. I think these are clearly recurrent. They're identifiable and many of them can be alleviated; and that's really why we're here.

The first of these, I think, is the social problem that everyone runs into. The problem of astigmatization of people who behave unusually is a very, very old one. Just last week on one of our travelling clinics I was getting a family history from someone and they told the old, old story of the grandfather coming to Colorado many years ago and being unable to buy food at the local store. The local ladies ganged up on him because they thought he was alcoholic and they persuaded the storekeeper not to sell him any food. And this still persists; there's no question of it. It persists on the current modern level in a more subtle form, in terms of getting jobs, keeping jobs, in terms of insurance, in terms of just daily living. This is a very, very real one and an unusual one. Not too many disorders seem to bring forth such a reaction from society.

The second one is the emotional aspects for the families themselves. We all hear and, I'm sure, know that the early stage of this disease, if it's not suspected, can really wreak havoc within a family. People don't understand the behavioral changes and this can really be a disaster for a marriage, for children, and so on. Later on, of course, as the disorder progresses, then, there's still other problems involving the whole family. And I think one of the things that troubles me most about it is the problems for the person who is at risk, who may not develop the disease but who has all of the major concerns about this. And this affects twice as many people, of course, as the disease itself; maybe still more.

In the past there has been a great tendency, of course, within families to hide this fact. This, I think, is changing but it's quite difficult because of the astigmatization problem that I've just mentioned. This often results in a real delayed explosion within the family when this information is withheld and then comes out later on.

Then the problem, of course, of medical care itself. This is, I think, from my standpoint, trying to steer families into optimal medical care is a difficult one and it's curious. The family physician, on the one hand, knows very little about this disorder; he's not equipped to handle it. On the other hand, the skilled neurologist, for some reason, doesn't want to see patients with this problem. I think he is scared of what is going to happen if he makes an early diagnosis and feels, perhaps, he doesn't have too much to offer. But we actually have great difficulty getting neurologists to see not only people who are sick but people who are well.

I feel it's very important that someone who is at risk for this disease have a good neurologic exam. If he is young there is about a 99-percent chance it's going to be a normal exam and it's going to be very reassuring--you don't have any problems right now. But the neurologist doesn't seem to see it this way. So, obviously, there's a great need for education of neurologists. If we're all to be working in the same direction we have to communicate and the neurologist has to understand that we're here to help on a long-term basis and this is a long-term problem, even for those who are at risk. It just doesn't come and disappear overnight.

What about social services? I think the families I have talked with have all expressed the feeling that there's nowhere to turn; no one is around who can really help me get the financial assistance, to help me get out the community resources; there's no number to call. The Huntington's chorea organization here is helpful and I'm sure their experience is very valuable but I think more can clearly be done. Similarly, no one has expressed to me the feeling that there's

good chronic care facilities; the nursing homes rarely seem to work out; the state facilities for mental health seem also not to be appropriate. So, clearly, more needs to be done there.

We're dealing with a very important economic aspect of it too. If the family stays home to care for someone, who works? Clearly, we can do more.

In the West we have a very special problem and I should address this too, I think. The distances are great; there's a lot of people out here but they're far apart. The medical facilities are spread thin. We have to travel, families have to travel. There's relatively few concentrated up-to-date medical facilities. Clearly, what is needed is to hook into our current travel program all the possible things for such families that we can.

Well, with this background, I would like, at least, to go on to try to make a few suggestions about practical things that can be done; many of them not expensive, but I think all of them very helpful. Obviously, these are broad since the problems are broad.

The first, I would say, is that of education, and this needs to begin in high school. Fortunately, this is a time where educators are very receptive to the idea of teaching biology through human genetics. It's much more interesting to think about people than it is to think about drosophila -- no question of it. We have the facilities to do this, there's the interest, and you'll hear more of this today. It's got to begin there if the whole question of inherited disorders is to be brought in the open and everyone is to get used to this idea that we have certain risks and certain problems.

Then, of course, the big one is how to improve the medical care. Oh, excuse me, I should go back and say that there's a great need also for education of M.D.'s. I've already mentioned that the neurologist and the family practitioner don't know much about this disease, but also, the medical school is going in the wrong direction. Here, for example, we now have fewer hours to teach genetics and they've dropped the requirement to have a course in genetics before you get in medical school. We're going in the wrong direction; so it ranges from top to bottom.

Now, the medical care it seems to me that there's several things that can be done in 1977. The first and simplest is to have a phone number with a professional at the other end for, at least, a large area. This would be some place to turn. It could be a social worker, it could be a geneticist assistant; it doesn't matter. It's a person who would accumulate experience as to what worked, where to get help, where to go to get the community resources, which doctors were helpful, and so on. I think this could be done for a group of disorders, including Huntington's disease, and it would be very helpful for the families.

Ideally, I think we should be able to develop a team with a special interest in medical centers in this disorder and related disorders. What form this would take would depend, I'm sure, on the medical school. But it needs to be done in medical schools; it needs a geneticist; it needs a neurologist; it needs people working in research.

The question of legislation, I think, and the legal aspect of it, needs addressing. We all have special risks. It happens in families with Huntington's that this has been identified, and to endanger someone's job because he's at risk for a disease he may never develop is clearly wrong. Whether this goes the route of cases in court or legislation, I don't know; perhaps the first route is the one to take in someone who clearly has been treated unfairly.

The question of research, finally. In teaching the medical students, I use the figure of about 10 percent of people having significant inherited problems involving the central nervous system; and I think this is a pretty hard figure for disorders that are either clearly inherited or have a major genetic component. This is a lot of people with problems that are there for most of their lives.

Funds for this type of research are very difficult to get. We have to work with people. The brain is complex and we can't study it too well in *Drosophila*. It's expensive research and it's difficult to get funds for this. There's no question that supporting this area of research is one way to go to help all of the families with this disorder and many others.

Thank you.

N. WEXLER: I think your descriptions of the problems are succinct and your recommendations are excellent. I'm wondering about the implementation and wonder if it might be possible to have an ombudsman or some representative of a category of genetic or genetic-neurological diseases who might, in fact, be attached to Dr. Ferguson's program. Specifically, one of the questions that we've been hearing, or one of the problems that we've been hearing continually is that patients and families don't know what programs they are eligible for and they don't know what kinds of Federal, state and local programs exist for home health care, Medicaid, food stamps, whatever. And if it might be possible to have some ombudsman or some person that would be within the state health department who could somehow mend all of these fractured surfaces--be able to say, this is the network of services that will be applicable for these particular problems. And that person might also, then, work with the team at the medical school who could provide the genetic neurological research care. But I wonder--and maybe this is a question for both of you--if some kind of implementation of Dr. Lubs' suggestion might be worked out through the State Health Department?

LUBS: I don't think it matters which place it comes from. The important point is that there is somebody. This could be part of the overall genetics team; it could be part of the State Health Department. Provided there's communication, I don't think it matters. The main thing is that there's an identifiable person to help.

FERGUSON: I would agree. It doesn't make any difference where it is, and certainly, as you well know, there are precedents for this kind of cooperative activity already.

N. WEXLER: It may not make any difference where it is, but what matters is that it is.

MCKHANN: Let me ask you a question about that because one of the things we'd like to be able to point to are places or models where this kind of thing is working already. Do you know of...where would you say, Herb, this is best being done?

LUBS: You all probably are better at answering that question than I am. I know it's not being done here; it's not being done in this part of the country. It's a very hard sort of salary to get. It's new and it's a type of care that isn't conventional. I'm not aware that this is being done anywhere. I suspect it is, but perhaps you all have heard of it.

GUTHRIE: It might be appropriate to comment at this time, then, that there is something going on in the State of Maryland which is a state-supported Commission for Hereditary Disorders. And it might be interesting for you, Dr. Ferguson, to know about that and perhaps we might be able to establish such a commission here. It is state funded; it is ongoing; it's in its third year. And I think it may not be everything that we would like, perhaps, in the way in which Nancy just described, but it certainly could be a beginning.

LI: Dr. Lubs, you mentioned the problems of education, and so forth. In what way a high school education on human genetics could possibly help...what kind of high school curriculum would you suggest that would bring any kind of a result at all?

LUBS: Well, I think there are specific things that are being done already. And the approach as it is generally here, I think, is through the general problem of human hereditary, not specifically Huntington's disease. For example, we have worked out a kit so that the high school students now can do a cariotype and arrange human chromosomes, so they get the idea of what human chromosomes are like. And this is used for teaching that aspect of biology, and very successfully.

I think pedigrees with Huntington's disease can be used to teach dominant inheritance, for example. And what one does is not only to teach biology, but to get across the idea that these are common problems; that most families have one thing or another there that is inherited; and to take this out of the range of the mysterious. I think anything is going to help.

MCKHANN: Thank you, Herb.

Dr. Chisholm, would you...

TESTIMONY OF
R. NEIL CHISHOLM, M.D.
ASSOCIATE PROFESSOR OF FAMILY MEDICINE
UNIVERSITY OF COLORADO

CHISHOLM: Thank you. I'm Dr. Neil Chisholm, a family physician who has practiced in Denver for a period of 25 years and I'm now (for the past several years) a teacher of family medicine at the University of Colorado Medical School. A lot of what I have to say has already been repeated by Dr. Lubs but perhaps a little of a personal touch, because I have had, in my lifetime, two patients with Huntington's, and I will give a brief description of those two patients.

In addition, I am now teaching young family physicians, and I think young family physicians really have a very big part in the treatment of the Huntington's problem. We have 24 residents in the Family Practice Department at the University of Colorado, and I consider the role of the family physician (the new family physician, in particular), a big part of our training is in counselling. And counselling, I think, is one of the biggest parts of the Huntington's problem.

So my interest stems from being a practitioner for many years and having seen a couple of problems, from being interested in teaching young family physicians to be compassionate and being capable of delivering and consulting with families and helping them with their chronic problems that are related to Huntington's chorea.

Also, I have recently made sort of a survey of colleagues of mine to see how prominent the problem is in Colorado. And this doesn't represent the problem in Colorado, in particular, but I have found out that the numbers of family physicians I have asked about their contacts with Huntington's during their career, about one out of five people have seen a Huntington's case in their practicing career. Now, of course, they aren't all in Colorado and some of them have been families that have moved, some of them have been contacts from other states, and so forth.

But in my personal case, the first case I saw was about 22 years ago, and I saw this case in a fairly far-advanced stage of the disease. It was a male about 50 years old who was really totally incapacitated by a far-advanced case of the disease. And, of course, my biggest problem was we were treating this patient at home until they moved to California, and it devastated the whole family. There was a wife, he was the father, and he was in his mid-50's in a far-advanced stage of the disease and really totally incapacitated. There were two children at home in their late teens, I believe. I'm sort of vague on this; I couldn't get the records and it's been, as I say, over 20 years ago. But I remember how devastating this problem was to the whole family. This was an active case, affecting the whole family, affecting the community and registering the devastating effect it has on the community and the family as a whole. Now these people finally got to a stage where the mother had to move to California to be around family because she couldn't cope with the problem anymore and she needed help from supporting family groups. And so at that time I lost contact with the family without knowing whether any of the siblings developed the disease or not, and I've had no contact further.

A little different case, about 10 years ago, I dealt with a family who were patients of mine. This was the son of a mother who had Huntington's in a fairly advanced stage, and she was in her 60's. And the son, I believe, at that time was about 27 or 28 and since that time...well, it's been several years ago and I guess he's about 38 or 39 now and has five or six children. The mother has subsequently died, and the son has not developed the disease. He has a sister who has not developed the disease, but of course they're at that age group where they are really concerned as to whether they might develop the disease or not.

And I might say the patients' parents had a death in each of these families too. So the families were concerned about their welfare and the possibility of them developing the disease. And it impressed upon me the scope of the problem and how many people are affected by just one case of Huntington's. So I thought, you know, a little personal description of some cases that I've been in direct contact with, a little mention of the magnitude of the problem as it seems to be, and then my particular interest in teaching family physicians in the area of counselling, being able to utilize the geneticist as they should be, to be able to use the neurologist.

I agree that neurologists are very reticent to see chronic neurological problems, but I think the family physician has a good rapport with neurologists and if somebody can help get these patients with proper counselling and proper diagnostic techniques, that the family physician is probably the one that can do that.

So that, really, is the reason I asked to testify. And I've had a little more recent interest because of dealing with young physicians who are going to go out in the world and, hopefully, deal with this problem.

I thought I would just mention and list the problems, as I see them. I've gone over some of them and Dr. Lubs has gone over all of them, I think. But the original diagnosis, probably not so much a problem as it used to be 25 years ago when I first saw my first case. But I was a little confused about that and, as Dr. Lubs says, family physicians don't know much about the disease. And I think, really, internists and not many people do know much about the disease because it isn't all that common and they haven't taken the trouble to delve into it. And so, often, when they have a case, it's referred immediately and they lose contact with it.

One of the biggest problems that I see, of course, is the diagnosis in children not being possible before they are of marriage age and before they begin to have children, and the devastating results that occur if they are not properly counselled in this stage of their life.

And then, of course, the major psychological problems that occur in the entire family group and the neighbors and the community, that result from a case of Huntington's.

The social and economic disaster that occurs in the long-term care of a Huntington's practically devastates the family. The first family that I mentioned was economically and socially devastated by this one case and, as I say, they moved to California to be near relatives and friends that could help out with the problem.

Proper facilities, really, I don't know whether any place has ideal facilities to care for the problem. But certainly in Colorado we need increased facilities and increased understanding of the long-term care of these patients. The ones that I'm most familiar with at the present time are being cared for at the Veterans Administration Hospital on an outpatient basis and at Ft. Lyon Veterans Hospital on an inpatient basis. And that, of course, just includes those people who are veterans and the other people are left to sort of go it on their own and find help from social workers and state welfare, and so forth.

And then I think the other problem that we've all touched on is that community education is an important problem, too, because with any chronic devastating disease like this, it's so important that the community as a whole understands it, as well as those intimately concerned with it. If the community as a whole understands the problem of Huntington's chorea, it makes it much easier on the family that's directly confronted with the problem.

I think that's all I have to add.

MCKHANN: Let me ask you a question, Dr. Chisholm, in terms of impact of your program because this varies very much region by region around the country. What percent of the graduates of the University of Colorado go into family practice?

CHISHOLM: Well, I think we had 28 out of 125 this year.

MCKHANN: So it's about 25 percent, roughly?

CHISHOLM: 25 percent, correct.

MCKHANN: And has this been a fairly constant figure or is that varying?

CHISHOLM: For the last three or four years it has been that high. It had a slump in the past decade and now has had a resurgence. But this is pretty much true throughout the country. The increase in family physicians is quite well documented in almost all states, except maybe some of the eastern states.

MCKHANN: Dr. Lubs, did you want to make a comment?

LUBS: Yes. I think Dr. Chisholm has hit on a very important point. The emphasis in medical teaching and delivery of care today is on what is called "primary care". In applying for money to foundations for genetic disorders and for counselling, we are told that this is not primary care; this is secondary or tertiary care or something else. Clearly, this is primary care, and the route you have outlined is certainly the first way to go.

I think the Commission can do a great service by trying to identify all of this as part of primary care, not some esoteric, tertiary ivory-tower occupation; it isn't.

GUTHRIE: Dr. Chisholm, I think that perhaps the reason you have seen so few patients is because the families were hiding. And I think that...I see you're shaking your head... and I think it's important for the record. I want them to state that there has been a change of feeling amongst families that they've begun to understand that you cannot do research on ghosts; that you have to do research and care on real people. And I've travelled around this country with the Committee to Combat Huntington's Disease finding families everywhere. And I think that if the people in the community knew that you would like to see a patient perhaps come and visit you early in the disease, not at the end of the disease, and perhaps be more helpful to you and to your students, maybe more of your family physicians will know and see more Huntington's disease if they know that you're interested in seeing them.

CHISHOLM: Well, I couldn't agree with you more. And that is (we really emphasize this in our training program) getting to know the family well enough so they're willing to confide in you and that there be people associated with the family who are willing to confide in you. I think that's a very important part of our training.

LI: Well, I was very happy to hear the test on the family practitioner. Now, tell me this, the first time you saw an HD patient that was 20 years ago. If you see a patient tomorrow again, from your point of view, what progress have we made in this 20 years? What would you do which is different from what you did 20 years ago?

CHISHOLM: Well, I think the only big change...on this first patient I had, it was just episodic care. When he got sick I went out to the house to see him and take care of his colds, and his pneumonias and anything that might happen. And I sat down and visited with the family and tried to console them, and so forth. But I think I would broaden the scope of my attention to Huntington's today with inviting all of the family in for consultation and counselling; get in contact with a good geneticist and have a neurologist; and it would be a team approach. And I think the family physician would be the one who would be the captain of the team.

N. WEXLER: We had heard testimony in some other cities that the requirements for the family practitioners, the curriculum requirements entailed less by way of genetics than neurology. Could you comment on that...that there are fewer courses in these areas?

CHISHOLM: Genetics is sadly lacking in family practice. In neurology, the American Board of Family Practice has realized the deficiency in neurology and is now becoming required rotation in family medicine residencies. This is at the graduate level, of course. So, we have recognized the deficiency in neurological training and are doing something about it.

N. WEXLER: Good. I take it that when you say "counselling", you mean more than genetic counselling?

CHISHOLM: Oh, definitely.

N. WEXLER: Do you give courses, then, in counselling techniques?

CHISHOLM: We do. We have an ongoing behavioral science curriculum. Throughout the three years of our residency program, behavioral sciences are a very important part on a day-to-day basis. So, counselling, from the family physician's standpoint, is more not genetics. I think the geneticists are more qualified to counsel in that area. But our counselling is with families in working out the problem as a whole.

MCKHANN: Thank you, gentlemen. I think we'll move on to our next group. On my list I have Sandy Keener, Dorothy Snyder and Bill Mayer.

Dr. Mayer, I understand you have some time problems here. Maybe you could just go ahead and start.

TESTIMONY OF
WILLIAM MAYER, M.D.
DIRECTOR, BIOLOGICAL SCIENCES CURRICULUM STUDY

MAYER: I'm very glad to have this opportunity. I speak from a bias that I should make clear initially; and that is, that my bias is educational. I'm very concerned that we have an educational program broad enough so that people perceive the problem.

In India, for example, we went to talk about birth control and we were going to solve their population problem. And the Indians, in essence, asked us, "What population problem?"

Now, we come to the same kinds of things when we deal with genetic disease. This is a simpatico group that feels about the same way, and we get to reinforcing one another, and begin to think that other people feel this way and perceive as we do; and they really don't. And the first thing that has to be done is acquaint people with the fact that there, indeed, is a problem.

I think it is absolutely essential that an educational program is, first, necessary if there is to be support for an action-oriented program of any kind. Now, we have conducted some preliminary assessments and our data indicate that education regarding genetic diseases, in general, and Huntington's disease, in particular, varies either from primitive to non-existent. Not only is this a general observation across the population, but in cases where one would expect it to be otherwise; that is, in the training of...(Incomplete due to change in tape)...particular, is at a fairly rudimentary level except in a few cases. When people who should be most informed are given such short shrift, it's little wonder that the materials we have for others are in limbo. For example, most texts that deal with disease still spend a majority of their time, if not the entirety of the book, on the coverage of long-existing disease categories such as respiratory diseases, food-and water-borne diseases, venereal diseases, with very little attention to the genetic diseases at all.

Now, with these observations, with this needs assessment, it seems proper that we should begin to think of a generalized educational program that could be developed with specific emphasis for

particular groups of people. I believe, for example, there needs be specific programs in the training of medical doctors concerning genetic diseases. All of the paramedical and support people should also be exposed to somewhat similar information. Now, as these will be the individuals who largely meet, diagnose, prescribe and predict regarding matters of disease and health, this training should include not only symptomology and diagnosis of treatment, but counseling as well.

Now, another group that needs this same kind of approach are the parents who may not be at risk but who are in need of counselling regarding children already born and whether to have more. The victims of genetic diseases, themselves, need an educational program as to how to cope with their situation. And finally, but by no means least, the citizenry, in general, needs an understanding of the problem and an empathy with it. It is unfortunate that most citizens have no concept of genetic diseases whatsoever, nor any ability to recognize people so afflicted, nor any sympathy with the problem.

In short, I hope it will be possible for the Congressional Commission on Huntington's Disease to have as one of its priority goals broad educational concerns across the spectrum of citizens who will contact, professionally or socially, individuals who are either threatened by the probability of genetic disease or are actual victims of it. I very strongly believe that without an appropriate educational program designed to meet the needs of the varying groups that will contact that segment of our population experiencing the trauma of genetic disease, there will be very little chance of a successful program that can effectively deal with this issue on its wide varieties of levels and in significant degrees of detail.

Thank you for the opportunity to read this into the record.

MCKHANN: May I ask a question, Dr. Mayer, and that is, whether you see the situation changing. I've had the impression that kids in high school today know a fair amount about DNA and what it does (much more than I did at that age, that's for sure--maybe now). But the question I'd like to ask is whether it's really as weak as you make it sound, not thinking about today's parents, but tomorrow's parents who are, say, in high school or below now?

MAYER: Everything is relative. I agree with you, the children are exposed to more information. But exposure to information is not the same as understanding a problem. Many children can tell you about DNA and the nucleotides, and so on, but we don't take that next step of the significance. There is a marked tendency on the part of schools to move away from anything they might feel controversial or socially oriented because of community pressures. If you want to play it safe, you play with only the details of a problem; that is,

you talk about the structure of DNA but you don't carry it to the next steps because we have all kinds of pressures there. For example, amniocentesis, which is a common technique that we all know about, we have been castigated for introducing this technique or discussions of this technique into schools. Now, we didn't think this was a problem; it was no problem to us originally. But in actual fact it was, because inferences were drawn on the part of the parents. For example, you are discussing amniocentesis, which is a technique of prenatal diagnosis. Now, the only reason you're discussing that is to determine if there is a problem with the unborn child. And if there is a problem with the unborn child, then abortion may be recommended, and we are against abortion and, therefore, take amniocentesis out of your classroom. We don't have openness in our classrooms in regards to these kinds of things and I think we need to have prominent focus. As I say, we give the child information, details, but we don't tell him what the significance of this is; and that's the important thing.

GUTHRIE: I just wanted to say that I'm very impressed with what you are saying because we have had to fight a battle to educate our people who believe that all we should do is spend our dollars on basic biomedical research only, and who don't understand the value of what you have said. And I hope that people will hear and that we can help you in what you are saying. I think it's excellent.

MAYER: Thank you.

LI: Dr. Mayer, since you make such a strong plea for education, I wonder if we can't extend it just one sentence more. If we have this educational program, does it follow, or doesn't it follow, that genetic counselling should be better understood and better accepted?

MAYER: By all means. I think it must follow. It isn't a matter of should it or shouldn't it; it must. This must be an accepted technique. We're in that stage now where genetic counsellors are thought of like psychiatrists were many years ago; you know, anybody who visits a psychiatrist should have his head examined. Well, we get into the same kinds of things with genetic counselling. There's still a stigma to doing this and that must be removed at all costs in the public mind.

GUTHRIE: Don't you think it would help, too, in removing the stigma, if we could get more families with all the genetic diseases to be concerned and come for help and work together.

MAYER: I think that's essential. I don't think...no matter how important Huntington's disease is, it's a part of a complex of other diseases which, as a whole, are ignored, although they have the same kind of root cause and they require the same kinds of viewpoints and treatments, and so on, and acceptances. I think it would be fatal to take only one of these and say this is the most important of all--Tay-Sachs, the whole group should come together.

N. WEXLER: You mentioned that there's little attention paid in high school texts to human genetics. I hope you don't take this question as a criticism. I know some of the best biological texts that are made and used in this country are from Biological Sciences Curriculum Study. I wonder if you could just comment on the amount of space in this text that is devoted to human genetics.

I know that you did a study that came out with these statistics and I don't know if you have them available to you or not. They were impressive when I heard them and I wonder if they could be put in the record.

MAYER: I don't have them at the tip of my tongue. Basically, the first problem you see in the textbook is an understanding of Mendelian genetics, and then maybe moving into population genetics, and then the structure of genetics. The human genetics, when you put this in as an additional item where...what are the human implications for this...something on Down's syndrome, perhaps, will be present; not much else. It is not that detailed in human genetics.

The concepts originally are to have the child appreciate that, indeed, he is a product of a kind of genetic inheritance that goes along. But the implications of that and the implications for humans...now, we're seeing a change. More and more, we're beginning to regard biology as less of a dissection of frogs and earthworms and more of an understanding of what's going on in humans, which really is going to be far more valuable to the average child and society. It's still pretty small; it's moving. But moving content of a curriculum is like moving a graveyard, and just for about the same reasons.

GUTHRIE: I have a very practical question. Where would you like to see the funding come from to support this kind of education?

MAYER: Well, I think it has to come from the Federal government. It will not come from local areas at all. The problem with American school systems (and it's a good problem--I don't mean this in a pejorative sense)...American education is very largely local. The local school boards have control of what is used and so on and these people are not in agreement among themselves as to the way education goes. In some communities it's one way, and some another. This has to be a Federal effort; if not Federal, it will not be done.

N. WEXLER: You seem to be talking about a general consciousness-raising.

MAYER: Yes.

N. WEXLER: Do you have any other recommendations for how this can be done other than through grade school curricula and even continuing medical education and allied health professionals?

MAYER: Oh, yes. I regard education as an extremely broad topic. I don't regard education as something that takes place only in a school classroom. I think television, very strong; motion pictures; community programs. You can't just reach the oncoming population; you've got to reach the existing population. There's a tremendous adult population now that has simply had all of this material bypassed; they just don't know. And they are very important to reach.

N. WEXLER: And educational programs, in those media, are generally privately supported?

MAYER: Yes, for the most part. And I think we could get private support; I think we could also get broader support from some of the agencies that put money into educational television--U.S. Office of Education, for example.

MCKHANN: Mrs. Keener, would you speak next?

TESTIMONY OF
SANDY KEENER
ARVADA, COLORADO

KEENER: Yes. I'm the other side and this is probably a very good time for me to testify because I think I've spent the last two weeks in constant tears.

My mother is presently, I would say, dying of Huntington's chorea. She has the disease and that makes me a risk person. I'd like to explain a few of the anxieties, fears and frustrations I've experienced since I found out that my mother had HD. The pressure is so impossible to explain unless you've lived through it. There are no words that can explain when you feel like you're drowning and you don't know where to go for help or where to turn.

We learned that my mother had HD about a year ago, after my aunt's condition was diagnosed in South Carolina. I think for a few days I lived in total horror, unable to sleep, work or think. I've learned more about the disease from the literature distributed, through Mr. Wilson and his Council, and through Marjorie Guthrie and her Council. After I learned of this, I just slowly began losing interest in life.

My mother had several years of severe problems. She was diagnosed as everything from being chronically depressed to the change of life, which seems to be a good cause for most things nowadays. When I was in my high school years I watched her change from a loving, giving mother into one that didn't care and was very hateful. Her smiling face became yells and screams, along with nasty comments and temper tantrums. She became capable of destroying a complete

living room, breaking the windows, throwing the lamps, throwing anything that was in reach, and finally, taking a knife and splitting the furniture. This, of course, was diagnosed as the change of life. Then came the neglect of not caring for her family, withdrawing from friends. Slowly came the inability to cook, clean or drive; along with this increased the temper tantrums, slightly slurred speech and motor ability problems.

After trips to the doctor and a stay at a mental hospital, she was diagnosed as chronically depressed and put on Thorazine. As the symptoms progressed, everyone kept silent trying to ignore the fact that something was terribly wrong. Everyone would say slightly, "She's mentally ill; let's just ignore it" and would let it go by.

Upon my aunt's diagnosis and because of the chronic behavior problems, she was again admitted to a mental hospital in Colorado Springs. She was completely out of control at the time, yelling obscene language and accusing every person in the room of every hateful thing she could think of. Her legs were flying high, her arms were jerking, and there was anguish on her face. She was allowed to return home until she became uncontrollable. My stepfather chose to ignore the problem and keep her home. I think this was what he felt was the best thing to do. He refused to listen to anything I had to say that I had learned on it and he wouldn't read the literature put out by the Council. The doctor, I basically blame, because he just was not familiar with the problem. He had never had a Huntington's patient. He got out his little books and read on it and that was about as far as we got.

I can honestly say this last year for me has been living hell; not only for me, but for my family. My mother has been asked to leave public places because the officials thought she was intoxicated or drugged. She couldn't eat in public places because she throws food in a three-foot area around her. She gets her tongue twisted and chokes, spitting up everything that is inside her. She has grease and other food substances all over her face and hands, especially when she can't hit her mouth. Every few minutes she has to go to the bathroom and because of accidents she has to leave her pants unzipped. Many times she is unaware of her pants and has walked outside with only her underpants on. She is unable to put on her undergarments or dial the telephone. This has resulted in excessive toll.

Do you know the anguish of hearing little children at play, running up and ringing the doorbell and then running and hiding, just so they can watch the "crazy lady"; or trying to get the nerve to walk up because the "crazy lady" has treats every day after school.

The television and the radio are not allowed to be turned up because they make her nervous. If you disagree with her she goes totally uncontrollable. She can't unwrap any bread, she can't hold

a cup of coffee, and she can't do the dishes. The personal care of her body and her teeth have been neglected for some time. She is losing teeth without even knowing it. She can't walk without stumbling and her daily walks outside had to be discontinued because the police picked her up for being intoxicated.

These are only a few of the problems. But what's really ironic is she hasn't been told that she's sick. The doctor in charge had seen a couple of cases and was basically unfamiliar with the disease. He did his best but it left you with a very helpless feeling.

The major problems we have run into are the lack of knowledge from the doctors, the lack of understanding about the disease, the future diagnosis of the disease at an early age for the patient, where you will put a member of your family for the duration of their life when the time comes; and, again, the good old subject of money ...where can you afford to put her? You can talk to the genetic counsellors at CU, but to get started that takes \$40 and what happens when you don't have \$40? You turn to good old Mr. Wilson with the tears.

After my mother fell several times and had other numerous incidents, we decided to start looking for a home or the equivalent. But where and what next?

It took a suicide attempt three weeks ago to get into the hospital without a court order and all the red tape involved. We still aren't sure about her destination after all the tests are completed. Money is running out and the insurance payments are small in a private mental hospital. She can't come home because there is no one to watch her and she is destructive to property and herself. My stepfather retires soon, so what next?

He is also, at this point, ignoring the problems. I asked him today to testify or just come to listen and he just couldn't.

As for my point of view at being a risk person...and having to deal with my mother has increased the feelings of helplessness and increased the pressure on myself and my family. It has affected my outlook on life, my attitude toward my child and my husband, and it has destroyed confidence regarding my ability to function on a job. I'm an Executive Secretary at Mountain Bell and you would think I was a clerk straight off the street with no education. I told my boss of the condition because I've had to call off work several times to go to Colorado Springs and now I feel as if my boss is treating me like I have it. He's watching my moves and my work ability, and I just feel it's at the end.

I can't put the fears and frustrations into words because it's lying under the surface. I tried hard to accept it and to adjust but I've only managed to try to hide it. I am learning to exist with the knowledge. I live one day at a time but it's in total horror. I look at my child and husband and feel guilt. I don't want to plan for a new home or extend too much money because I fear the financial burdens that my husband may have to face will be totally unbelievable at the time. I constantly watch myself for anything that looks odd: a slight stumble, or if I have to repeat myself, a typing error, a decrease in typing speed, dropping a glass in the kitchen. These are normal failures, but I ask myself, "Am I all right?"

The unknown is deadly. I think it's a stress that could almost kill anyone. How can I ask any man or child to love me when they can look at what they may face in the future. All of the above are playing a heavy toll on my life and my family's. And I am asking for not only my family, but for all the other HD families, that we please have some assistance.

I feel that counselling is needed for the entire family; doctors are needed to become familiar with the disease; and more research needs to be done for diagnostic purposes, plus the cure. Most of all, I feel financial assistance or where we can go for help is needed to be known. I also feel that we need an adequate place to put patients.

This morning I spent the first hour talking to nursing home and to some social service people, finding out what we could do with my mother. We have to decide by Saturday. Many nursing homes, I have found, in the Denver area run from \$600 to \$800 a month. I don't know if she will qualify for Medicare or social services; I'll have to find that out this afternoon. My stepfather was supposed to do this but he told me last night he can't. He's just too upset to do it.

Right now, I think I'm united with probably everyone in this room that's connected in asking for help of any kind. My husband was to be here today to testify but he was called out of town to New York and his testimony is outside in the hall...because it's affected my marriage. I have a wonderful husband and a wonderful family but I have just totally withdrawn from everybody.

GUTHRIE: Sandy, I have to reach out to you and tell you that you are not alone, and all you have to do is to reach out... there's a lady sitting right next to you who, herself, has been a helping person, a devoted person, and wants very much in the organizations of the families with Huntington's disease to organize and help. And the first step, once you get over the shock, is to start doing something yourself. And if you get involved...you're healthy

and well today and you may never have Huntington's disease. So, your job is to start working. And you reach out to Dorothy over there and let her hold your hand.

MCKHANN: Of what you've done, what has been, so far, most helpful to you, because there may be other people who would like to know how to start.

KEENER: I'm still at the point that I don't know how to start. As of last night, we're still running in circles. I learned of Dorothy, here, through a friend of my husband's from CU. He was an engineering student at CU, and through her we learned of Mr. Wilson. I can honestly say Mr. Wilson has been my salvation at the time I talked to him and his daughter. But I honestly can't say right now I've gotten any help from any place on anything.

I was having several symptoms and I was put in the hospital. This was a month prior to the fact that we found out my mother had this disease. And they diagnosed that I had hypoglycemia. Well, at that time, I even tried to see doctors to see if maybe I could have it. And the first neurologist that I went to, he almost told me that that was absurd, you know. You don't get it; it's a rare known disease. And he actually told me he didn't know anything about it. So, from there I went to Dr. [Sneck] at CU and he examined me and told me that I was fine. But the fear is still there and every day as we're looking at my mother and dealing with my mother...I say right now, financially, money is running out, my stepfather is ignoring it, I'm put in the position right now of trying to make the decision of what to do with her; and this is a human being's life. I mean, she can't control herself and she's having trouble functioning, but I don't feel that I have the right to say, "All right, you have to go in a home for the rest of your life." Whereas, you know, when I ask for money...like, this morning...most nursing homes, they don't want a Huntington's chorea patient. They're too much trouble. I found one today out off of West Colfax that would take her for \$600 a month. But now I don't know where the money is going to come from, you know, to pay her. And right now she's in Brady Hospital in Colorado Springs and that's costing an extreme fortune. And right now, I can say, we're not getting help from anywhere; you know, I just am very much at a loss right now and I'm just drowning. Then, I look at her and I know what she's been through and I think it's so silly.

GUTHRIE: No, it isn't silly. And it's important that you hear what Dorothy has to say.

MCKHANN: Mrs. Snyder.

TESTIMONY OF
DOROTHY SNYDER
BOULDER, COLORADO

SNYDER: My experience with Huntington's has dated back 34 years, when I married into a family that had it and I didn't know it and had two children. We had a great deal of trouble getting a diagnosis, several years. In 1947, although we had learned at that time what he had, we decided to go to Mayo's and I think you would be interested to hear that the head of the Neurology Department of Mayo's, in 1947, asked my husband if he would walk up and down the hall so that his staff members might observe him. He didn't think that any of the neurologists in that department had ever seen a Huntington's.

He continued to work in a professional capacity until about the last three years of his life, when I said, "Let's stop working. It's too hard for you to compete with well people." And it became easier then. He stayed in the home until the last six months of his life. I had to go back to work and it was difficult finding a place that would care for him. We did find a nursing home. I'll back up just a minute. He was a veteran; we tried to find a Veterans Hospital facility of any kind that would take him. They would only let him come for a few days, or a week or so, and then we had to find another place. This has changed today, I understand, and Veterans Hospitals have Huntington's people, although it is not a service-connected disability.

The nursing home where we finally got him in kept him for a few months and then said they couldn't care for him anymore. We would have to have him committed to the state hospital. The Veterans Hospital said, "We will take you back for a few days until you can make arrangements." And he died in the Veterans Hospital in 1957.

During these intervening years I wrote to everybody that I had leads to all over the world. There must be some good that will come out of this somewhere; there must be some kind of research that will stop this. I was very sensitive to the fact that I had two children; a feeling of guilt about it; a feeling of great resentment to my husband's family who had not talked about it, from whom I could get no information.

Mayo's came out with a little booklet, "Huntington's Chorea in Your Family", by [John S. Pierson], which I finally got hold of, I think, through the Veterans Administration Hospital here in Denver. I felt that this was about the best thing that I had learned and used it when I informed my two boys about the disease. And I felt very sure by the time they reached that age of 35 or 40 we'd have a breakthrough.

In 1967, I was devastated to find out that my younger son was showing symptoms. I was sure that this was psychological because he had been informed, he had an open mind and was reading all he could get hold of. He had a good physical examination and the doctor said, "There's nothing wrong with him but this disease is so rare I don't feel competent. Please take him to a neurologist." We did and the neurologist said, "I'll tell him he doesn't have it. He can't handle it if he does."

Well, the boy was 20 years old at that time; he didn't wait until he was 40 to show symptoms. He had already had two years of university. He took two more but had to give up 15 hours short of his degree. He was training in sociology. He came back home very depressed. Through good friends he got into a cerebral palsy center and did volunteer work for five years. Incidentally, I feel that there's a compatability in these two diseases. And if there should be health care centers set up some time, I think these two diseases could work quite well together. This was therapy for him; it was good for me. And from all the repercussions, it was good for those little kids he worked with. The love and devotion that they showed him I'll always remember and treasure. And he got over the depression. He found out that there were other people in the world who had problems and he became a contributing member of society, even in his disability.

He commuted by public transportation about 70 miles a day. We would drive down from our mountain home and he would catch a 7:00 a.m. bus out of Boulder to come into Denver, change buses. And he did this for about 4½ or 5 years until he began to fall. I said to the doctor, "It's too dangerous." And the doctor said, "Let him go on as long as he can." And we did, and finally he just gradually realized that he couldn't do it anymore. So, he began staying home by himself. I had to work and this became of great concern to me. And when he began falling at home and needing eight stitches to sew up a chin, things like this, we knew that I couldn't give him the care that he deserved any longer.

I feel that I have been one of those fortunate few who found a wonderful place for Dick to be cared for, the Good Samaritan Health Care Center in Boulder. It about killed me when I placed him there but, thank God, it was there. They loved him, the doctors, the nurses, the aides, the orderlies, the people that swept the floor. He developed a rapport with them that was beautiful. He had pillow fights with them and I was afraid he might get thrown out. He knew a sprinkling of different languages and could talk to the hired help in a few different languages, and they loved it. He was a good sport, he was cheerful, he had company.

When he finally gave up and realized that there wasn't going to be divine intervention or a medical breakthrough, I feel, in love

for his mother and just general good sense, on July 15th, last year, he stopped eating. And I mean he stopped eating; and he lasted two and a half months. I consider that his life was not in vain; neither do I think it is fair to bring people into the world that have only a chance.

My great concern has been and is basic genetic research. At the time I went into the Biophysics Department of CU Medical School, I begged them to do this. And they said it's too rare a disease; we can't take the taxpayers' money for it. I think we've proved that it is not so rare. And that to me is the primary thing that we should consider, one way or another. We have people interested in doing basic genetic research now. Let's give them the money.

MCKHANN: I'd like to, if I could for a moment, pick up on the last point that Mrs. Snyder raised; and that is, just to mention a few words about the problems of those in research right now. At the Federal level the supportive research really is in greater jeopardy now than it's been any time that I've been associated with biomedical research. And many of us who are involved in this are at a loss to know what to do because there is an anti-intellectual climate almost taking place. And it's important, I think, that people like yourself continue to try and get this message across, particularly to people who make these decisions, both at the Federal and state level. We're doing what we can in this regard, but I think you do too.

SNYDER: I've tried for 34 years.

GUTHRIE: But, Dorothy, you agree, as you say, we know now that this disease is not as rare as anyone believed. We're still finding more and more families. I should say publicly that with a lot of publicity that we had just recently, in the last month, the Committee to Combat Huntington's Disease had 179 new families come to us. Now, that's a startling figure when you think that we've been around for nine years and our average used to be 33. So, yes, we're finding the families, but don't you also feel that the families should work together to be the constituency that can fight for that funding that you're speaking of? We cannot do it alone, can we?

SNYDER: No, that's right; it's a problem.

WEXLER: I think one of the really important things about today also is that some of those same people that you once talked to so long ago are now here listening to this testimony and involved in research on Huntington's, not only because there are more Huntington's patients, but because I think that it's becoming more and more clear that the answers to Huntington's disease might lead to the answers in many other diseases as well; and that it isn't only our own population that we have to increase.

I have one question about care, and that is that it seemed that your son's involvement working with children who also had a disability gave him so much. I wonder often how much kind of devastation that happens with Huntington's is really due to the kinds of care facilities that are available to them. And if you feel that your son's maintaining an active life and involvement with these children may have in some ways even affected the course and progression and severity of his disease. Do you feel that that might have just even altered the way that the disease affected him?

SNYDER: I think it affected his acceptance of it, realizing that there were other people suffering in the world. I think he saw firsthand the value of various kinds of therapy. It made him more willing to experiment with therapy. This is my feeling. I don't know if it's scientific. I doubt if it in any way affected the progression of the disease.

WEXLER: If there were care facilities for HD patients, do you think it would important to have people with other disabilities included, not just a care facility for HD patients alone?

SNYDER: Yes, I do. I feel that there is too much hopelessness connected with the disease to isolate it. I think it could better go along with something else where people can see how devastating other diseases can be too; and that, perhaps, they can work together.

WEXLER: I just wanted to say, you have my great respect.

GUTHRIE: And I want to say for the record that I met your son, you know, when I came and I thought he was beautiful. And his giving to those young children, I think, did shine in his eyes. He had that quality; he was beautiful.

MCKHANN: Dr. Kimberling, I was reminded to ask you to please say your name and affiliation before you start.

TESTIMONY OF
WILLIAM KIMBERLING, M.D.
ASSISTANT PROFESSOR OF PEDIATRICS
UNIVERSITY OF COLORADO

KIMBERLING: I'm Dr. Kimberling, Assistant Professor at the University of Colorado in the Department of Pediatrics. And I have been involved now for a year and a half to two years in genetic research into Huntington's disease and in the course of that research we have collected information on about 200 families and we're continually collecting more.

I have deliberated for quite a long time to decide exactly what I'm going to say up here, and I would like to cover a few points.

The first has to do with exactly what Nancy Wexler mentioned before, the relevance of Huntington's disease to everyone in the population. It's certainly difficult to arouse public interest in the plight of families who have Huntington's disease. In fact, it's natural, and people are repulsed and avoid contact with these families. This is something that's overcome by education, and if you work hard at it, it can be done. However, the problem still remains and it's difficult to have a cute poster child, like they do for muscular dystrophy or hemophilia to arouse sympathy in the public and motivate them to do humanitarian acts because Huntington's disease is not a cute disease; it's a very tragic one as we've heard here. I find it particularly tragic that despite the fact that people in the population know about hemophilia and muscular dystrophy, I believe Huntington's disease to be more frequent than those diseases and probably more frequent than both of them put together. And yet, I know very few people among my own acquaintances who knew about Huntington's disease.

Now, one would hope that a humanitarian appeal might induce the public to provide the funds necessary to do what needs to be done in order to provide a cure and to provide care for those families with Huntington's disease. I don't believe that this is sufficient. Even though we might quote what a geneticist considers to be a high frequency, let's say one in a thousand, this is not sufficient; and legislators will say, "Well, why? We should be giving the funding to other diseases such as heart disease or cancer. A great number of people are affected by those diseases." So, it's only natural that funding for Huntington's disease would tend to take a back seat to these other problems. However, it can be shown that significant progress in research into Huntington's disease could very likely benefit every individual in the population.

As I mentioned before, millions are being spent for research to find cures for cancer and heart disease, and these are both laudable and necessary goals. However, if cures for these diseases are found and our life expectancy is significantly increased, we will be condemning a significant proportion of our population to spending their final years in homes unable to care for themselves and unable to think with the same efficiency with which those of us think in our middle and early years. I find this particularly tragic. It seems to me that the government is not thinking ahead. We're not thinking of the consequences of the cures of these more common diseases in relationship to what the effects are going to be on our population. The mean age of our population is increasing every year, the baby boom has passed and we're now seeing those babies in their middle years; and this is going to reflect in a great burden in the future.

My point is simply this: that Huntington's disease is classified as a presenile dementia. It has many of the characteristics of aging and many of the symptoms that will occur to all of us as we

reach advancing years. Certainly, a greater in-depth knowledge about Huntington's disease can increase our own ability to understand the aging process in each and every one of us. And significant advances into the area of treatment and cure of Huntington's disease is bound to affect each and every one of us and could possibly lead all of us to have a better quality of life in those years that remain before us. This is a very important point and a great deal of public education is going to have to be made in order to alert the public to the fact that Huntington's disease, while it is very tragic to certain families, and while it is very rare, can be used as a model, a very important model, to allow us to understand basic biological processes in the genetic control of aging.

Very much the same thing could be said about Huntington's disease and its relationship to mental disease. In-depth knowledge about what causes the psychological problems that occur in the early stages of Huntington's disease could very well provide the clue that would allow us to better treat more common diseases such as schizophrenia. This also is quite important and could alleviate a considerable burden to the population.

That's basically the core of my comments with regard to why I think the government and the public at large should spend a considerable amount of their money and their time and efforts at helping these families with Huntington's disease.

There are other more specific problems concerning research that I would like to point out. These are specific problems that I've run into in my own study with Huntington's disease. One of the most glaring of these is that our funding is provided for a more basic kind of research project and we study several diseases of which Huntington's disease is only one part. All of our efforts are oriented towards answering specific research questions. As such, we have very little time available in order to help families where the help is needed, and we feel very guilty about this because we're uncovering new cases, new families, and bringing to light the problems that surround Huntington's disease; and after doing that we're unable to do very little in the way of helping these families. We have no one that we can refer them to, no one who can help them sufficiently. We refer them to Mr. Wilson and he helps a lot; however, this is not sufficient. We need help at the governmental level, social workers, people who would know where these people could get economic help, who can get medical help, and whatever else that's needed.

The geneticist is indeed a focus for many kinds of genetic problems because, even though we are doing research, we're very much committed to uncovering and finding these cases and we attack this problem vigorously. And by doing so, we create a problem for ourselves because we have to find something to do with these families. And unless funding is allocated to allow us to refer these families to

appropriate agencies and that those agencies have enough money to care for these families, there's little else that we can do for them. And I feel very guilty about not being able to help the families more than we have been able to do in the past.

There's a great need, I would like to say, for in-depth population studies on Huntington's disease and other neurodegenerative disorders. Even though we've collected quite a few families, we really don't have a very good idea about frequency of Huntington's disease; either the frequency of affected individuals or the frequency of individuals at risk, or the frequency of the gene in the population. This is important from the point of view of planning. As you've already mentioned before, it seems like this disease is more common than previously supposed. I believe that this is true as well. We had about six months ago, approximately 120 families, and at that time I said, "Well, gee, we couldn't get any more in this state; this is a small state. It only has a couple of million." And yet, families keep coming out of the woodwork and new cases are popping up, it seems like, every week. And at the present time, we have over, as I mentioned before, over 200 families, and they're still coming. And so, the frequency of the disease, I've just roughly estimated, in Colorado, must be approximately one in 10,000, if not higher. And that's the frequency of the disease, and if you added the individuals who are at risk, the number of people who are affected would be even greater. So, it is not an uncommon disease, and compared to other kinds of diseases where a considerable amount of funding has already been allocated, Huntington's disease is by no means a minor problem. It is a major problem for the population. And it is certainly as great as phenylketonuria or Tay-Sachs or sickle-cell anemia, because of the devastating effects that it has on the family because it affects not only those individuals who are affected and who do carry the gene, but their relatives and their neighbors as well.

GUTHRIE: So, essentially, you could say that you could take your 200 families and multiply it by maybe a spouse and two or three children, and that would be a figure, right now, of people living who are affected with this disorder, right?

KIMBERLING: That's correct.

MCKHANN: I'd like to ask you, Dr. Kimberling, of an area you haven't brought up, but probably have run into, that we've discussed in our Commission meetings. You've mentioned your desire to get genetic data and we've had some discussions about the problems of personal privacy and how one handles this data. I'm sure you've run into this too. I was wondering if you could make any comments on that particular issue.

KIMBERLING: Well, there are several things that I would like to see done. Some of these can be implemented at a personal level and we do it. We take all steps that are possible in order to assure privacy. And in our research there are many people who would like to participate and to a certain extent are afraid of doing so because they're afraid that their records may be used by insurance companies or others. Our research, we maintain two classes of records. One is considered part of the patient chart, and this is subject to the same laws that any other bit of medical information would be that's entered into the medical chart. And, indeed, we can generate computerized listings, and this sort of thing, of many of our families. And even though we do this, we consider all of this basic medical information and it cannot be released without the patient's consent. And we take every step we can in order to assure their privacy.

At the same time, I think that states, or perhaps the Federal government, are going to have to address themselves to the problem of data that is collected in a purely research environment. And I don't believe that such records should ever be subpoenaable (sic) or should ever be taken out of a scientist's office. This is, in a way, even more sacred than information that's given to one's family doctor. This is information for the researcher...well, let me backtrack and say, the researcher, even more than the physician who works daily with patients, has to be absolutely correct in the conclusions that he makes, because even though it's not apparent, a researcher who says, "This is true" affects thousands of people. He does this by affecting the physicians who are working on the primary care for these patients. And if he's incorrect, that error is going to affect lots of people. So, it's very important that he obtains the very best and very most correct information that's possible. And for that reason, the information that a patient gives to a scientist in confidence should never, under any conditions, be used against him. And I think it would be a good idea for some states, or perhaps the Federal government, to implement a law, like I believe they have in Canada, where research records cannot be subpoenaed and cannot be taken out of a scientist's office under any conditions.

GUTHRIE: I can't let you get away without believing that you're not doing enough. I know that Dr. Kimberling happens to be also the medical advisor to the Committee to Combat Huntington's Disease. And we know darn well that you are doing what you can to help, and we appreciate it.

KIMBERLING: Sometimes it makes you feel guilty because you feel like you never do enough.

LI: Dr. Kimberling, you mentioned that you feel guilty because you couldn't help the patients out, their families, and there are no agencies where you can refer them to. Tell me what kind of agencies you have in mind.

KIMBERLING: Well, what I'm going to say is,,,I'll limit it specifically to Huntington's disease but it would refer to all other kinds of neurodegenerative diseases as well. And I should say, also, that we're learning; just the very fact that you're here has helped me a great deal in learning where to go in order to get some help for some patients and where to refer them. And it's generated interest in other doctors who now we may be able to get some more help. So, whether or not any money comes out of this Commission, it's done a fairly good job already. I should say a very good job, not just fair. Excuse me, I always qualify everything I say; that way I'm never wrong.

As I see it, one of the major things that we need is to be able to refer, first of all, people who are affected, to M.D.'s who are qualified to treat the disease and give support to the family as they're going through the process of the degeneration of this individual over the course of the last 5 to 15 years of his life. And as it is now, I don't know of any neurologist that I can call who says, "Oh, I would be happy to see that patient." I can think of other rare diseases that I've dealt with and the doctors are totally delighted to see it because it's something unusual and all of us have a certain amount of curiosity. But with Huntington's disease this doesn't seem to be the case; they avoid it. They don't want to see the patient; the families are "difficult" to work with--and I put the word "difficult" in quotes because the difficulty is real. It's there and the problems are ongoing, families are concerned each and every time the patient takes another step downward; and this is some concern to them and they want to get some more information; they want to know about it. It takes a great deal of patience to work with families with Huntington's disease and there aren't very many neurologists who have that patience.

In addition to this we have problems in obtaining care for the people at risk. Just two weeks ago I saw an individual who was at risk for Huntington's disease. They were planning...the father was the one who was at risk. They believed, as had been mentioned here, that they should not have a child who has only a 50 percent chance. So, they were going to have artificial insemination, and in addition to that, the father wanted a vasectomy. Vasectomies, for most of us, aren't terribly expensive, but to an individual who has had a parent with Huntington's disease and has had to spend the amount of money that has gone into the care for these diseases and this economic deprivation has occurred in generation after generation, they aren't that well off. And he was wondering about whether or not he could afford something as simple as a vasectomy. Now, here's a person who wants to do something about Huntington's disease and may be prevented from doing it because there is no money to pay for this one simple operation. I mean, after all, India pays 10 rupees for it and we make people who actually need it pay for it. And there's another area. One could go down a list.

I think that people who are at risk for Huntington's disease and the people who are in the families can clarify better the need that they have. You've heard so eloquently so far, the very poor care, the very poor understanding, even, that the families receive. And what we need, more than anything else, is somebody to refer the families to who cares and who would treat them as people, and who will treat the family as a whole, and who will try very hard to do something. Obviously, we can't do everything; we can't cure the disease. But we can provide support.

GUTHRIE: I also want to comment, though, and I think it's important that there are so many physicians who don't know that there are experimental drugs that are helping the two major symptoms of Huntington's. And it's unfortunate that the families and the physicians do not know about this. So this would be something also that I would like you to remind physicians when you see them, or patients -- look for experimental medications and use them because it does help some people.

KIMBERLING: Well, the surprising thing that I've found so far, we routinely ask family members who their family doctor is, and Huntington's disease patients don't have family doctors. They've gone from one doctor to the next, to the next, to the next, and they just have no one that they can rely on.

N. WEXLER: Bill, do you ever refer families, people at risk, to psychologists, psychiatrists, social workers, for emotional counselling?

KIMBERLING: We try.

N. WEXLER: Can you find them, who know anything about the problem?

KIMBERLING: No, we can't. One psychologist has tried and said he would work with them, but I don't think that's going to work very well because in this state alone, if you wanted to provide adequate psychological counselling I think it would take one or two psychologists full time. And it would be, probably, a neurologist could work full time on them as well. Despite the fact that it's a rare disease, the patients and the families require so much in the way of care and help. A Huntington's disease patient is not someone that you can bring in your office, spend five minutes with, whack on the knee and send out. You've got to spend time with them because it's more than just treating the disease; it's, at the same time, you have to try and give them an understanding of what's going on. So that a lot of the psychological problems which spill over onto the other members of the family can be avoided.

N. WEXLER: You don't have anything like a hotline or a walk-in clinic, or anything like that available to families?

KIMBERLING: No, I think the couple closest hotlines are Dr. Wilson's phone and mine. And I've begun to realize what a problem this is for patients because I'm beginning...because of our genetic studies we're not committed to looking at people in this state alone. In fact, the state is where we begin studying the families but, of course, they're spread all over the United States. I've gotten calls from California, from New York, from the South, from individuals who want to know, "What can I do? Who should I go to?", because in those regions they just didn't know. In fact, one of them just lived almost around the block from Dave Cummings. I knew where to send him, and the others I didn't.

N. WEXLER: Just two direct quick questions. Aside from the privacy problems, would a registry of HD families be useful in your research?

KIMBERLING: Well, yes, it would be useful in our research. A registry, in my mind, the main use would be to contact families when new and important methods of treatment are found, rapidly. It's my feeling that if there is a cure for Huntington's disease, the cure will be a prevention. And that we would have to treat individuals who are at risk before they even develop any symptoms and maintain them on that for all their life. And so that would be an argument towards contacting individuals, or at least having the capacity to contact these individuals, as early as such a cure is found. We can't let such important information just slowly disseminate out because it could take several years and that may cause undue tragedy in some families.

N. WEXLER: Have you been able to adequately financially support your HD and other research?

KIMBERLING: We can do the research, but you should see my lab. It's done, I'd like to say, on a shoestring. There's a considerable amount of money that's spent, but a lot of this goes for other kinds of diseases. But at the same time the kind of research that I'm doing is expensive and it's hard to compete with other people, and so it has been hard to get adequate money. It hasn't been hard to get money. We've been funded by several different sources and some of it has been specifically for Huntington's disease. It's been about \$40,000 every two years, specifically, for HD.

N. WEXLER: That's from private sources, correct?

KIMBERLING: It's from private sources and one from public--a small one from a public circle.

N. WEXLER: Thank you.

MCKHANN: Mr. Wilson, would you like to...

TESTIMONY OF
SEWARD WILSON
PRESIDENT, ROCKY MOUNTAIN CHAPTER
COMMITTEE TO COMBAT HUNTINGTON'S DISEASE
DENVER, COLORADO

WILSON: I'm Seward Wilson, President of the Rocky Mountain Chapter of the Committee to Combat Huntington's Disease. I have on record, members of our organization, more than 200 families, and I know of approximately another 100 families throughout this state. They are in nursing homes, hospitals, VA hospitals, and other different medical facilities. We were organized in 1970, here, three Huntington's families and one social worker. So, you see, we have grown quite rapidly and we are getting more people every day. And one of the problems right now, though, is with the younger people. They don't want to be known because they might lose their home, or they can't get insurance, or many such problems as that.

Now, I've been acquainted with Huntington's disease since 1947. My mother-in-law was diagnosed about that time by Dr. Moore who was in the University of South Dakota, and he was working in Yankton at the state institution. I'm unfortunate in one way; I watched her deterioration, not daily, but, you know, I'd see her probably once a month and then at certain periods, maybe twice a year. But I was able to note the deterioration and Dr. Moore had sat down with me and my wife and explained Huntington's disease and what we could expect. Of course, we were unable to really understand what was going to happen and the phases of deterioration. But out of the family of five children, four of them had Huntington's disease. One committed suicide, and my mother-in-law, she was killed in a car wreck after she had had it for 12 years.

So, I discussed the situation with my wife many times, and even back before her mother died, there were moments of depression because she really understood the hereditary part of it. But her father was very good to her mother and very compassionate and took very good care of her. So at the time she was killed in a car accident, she was still walking and she had it for over 12 years.

Then, in my family--I have one step-daughter and a daughter. My daughter will testify later this afternoon and give you her idea of the things that have happened to her. The wife, it took us about four years to really get her officially diagnosed. In 1967 the Fitzsimmons Hospital--I had taken her out there and they wouldn't officially diagnose her, but there was one doctor out of about 15 in the Neurology Section that knew anything about Huntington's at all and he had had patients from New York previously. And it was 1969 before I got her officially diagnosed and that was through Fort Logan Mental Health Center.

One of the things I want to speak about is, we don't have care for Huntington's people at an early point, and we don't have anything to help the family out. Now, they begin to lose their speech; we haven't been able to get speech therapists. We don't have physical therapists to take care of that situation at an early stage, and especially when it's been in a family, and you know it's coming up, or have an idea when these first symptoms appear. So that is when we need help; we also need help to keep the family together. We need the in-service help so that the father or the mother--whoever is working--can go ahead and work and support the family.

Like my situation, I had to work a few more years in order to retire and be able to support the family like they should--so, I had to take my daughter out of high school. She was out three years, but, thank goodness, she has gone back, finished high school and completed about two years of college now.

But at that early stage is when we also need a great deal of help around the home; and we need somebody that we can talk to, some counselling. So, that's the reason I think we definitely need a very comprehensive planning for the genetic diseases. And people, where they can go down the line and talk to the neurologist and see them--the psychiatrist, and the therapist, and so forth--and this all could be done there, because as the disease progresses, they begin to lose their speech and things like that; they become very irritated over a period of time and you have to take them to a dozen different places to get just one job done.

I do want to say to my people, I'm always glad for somebody to call me because I like to...it relieves my tension, also, to talk about it with somebody and go over the problems. There is a great number of problems in almost every field that you can mention. From the social because, one at a time, after the wife began to stagger and stutter a little bit, all of her friends, one family at a time, would go away. And today--of course, she's had Huntington's now for 13 years--we just don't have anybody to come to see her. But I still take her out. I go to certain...I belong to veterans' organizations and I take her out and hold her while she dances once a week, and then we also go to bingo once a week. So, that takes a great deal of love and takes an awful lot of care and personal attention, of course. Somebody has to be with her all the time.

We had one real bad incident happen last year--last April, a year ago. I went to the general office at Fitzsimmons, and I left her in the car, and while I was at the office, she walked into the hospital, staggered and sat down into a soft seat and couldn't get out. This excited the guy at the information desk so he called Emergency. They came back there and got her and strapped her down. And when I couldn't find her in the car, of course, I checked on what was happening. I found out she was in Emergency and, of course, we

got her out of that right in a big hurry. But the sad thing about it was, she does wear a medic alert; it was on her, and they didn't know. They didn't know what was wrong with her, or anything. They didn't even make an effort and that was at Fitzsimmons General Hospital. Now, what happened then, it took four months to get her back to normal, because then she lost control of herself and she would--sometimes she would see a building that looked somewhat like that, why, she'd even try to get out of the car when we were driving along. So, it took from four to six months to get her back to normal and get her over that fear.

And so, that shows you that the education of the medical and the professional, as well as the laymen, that's what we must have and that's one of the reasons that the CCHD must be kept effective because they can receive, disseminate, and pass on to us and distribute to the professional and to the public as well, the information concerning this disease.

MCKHANN: Thank you. I would just like to make a personal remark because we've talked about various problems in our Commission. We've talked about the need for Federal support, and the need for big policy changes. But one of the things that's very clear is that there will never be a substitute for people like Mr. Wilson.

Miss Rutan, would you like to make...

TESTIMONY OF
JOAN RUTAN, RN
DIRECTOR OF NURSING
LAKEWOOD NURSING HOME

RUTAN: I'm Mrs. Joan Rutan. I'm an R.N. and the Director of Nurses at Lakewood Nursing Home.

We have a patient now at the home with Huntington's chorea who we admitted to our home from St. Anthony's Hospital in 1973. Now, as was stated before, this is a chronic, degenerative disease, and most patients do not live to the point of progressive debilitation that this patient is in. She's in a semi-fetal position; she's very, very emaciated. Her arms and legs are at the point now where they're very brittle movements and she has very, very--as I said, very emaciated and has very little flesh on her. She does not communicate at all with us. The only thing that we can really see is that, as was mentioned before, with a lot of tender loving care, and talking to her when we do feed her--which is another point to be brought out--that it is unusual for a patient in this condition to eat and drink as well as she does. She doesn't have a nasal gastric tube; we do not have to give her intravenouses; and we do take our time, and she does eat the food that is strained and she

takes all of her fluids. Now, the nutrition that we are giving her is keeping her--you would think, well, possibly that she would be able to have more weight. But because of the constant movements that she is going through, it is impossible for her to gain any weight. We do get her up in the chair often, two or three times a day. We turn her often, every one or two hours, because of the possibility of breakdown of her skin. We do have a doctor who has been ordering lab work on her and different tests. And we would like to have funding, if it could be available, to better understand the disease. And he has started late in this disease and wishes that he could have started sooner because maybe they could have picked up something with these tests.

As was stated before, Huntington's chorea is hereditary and is a genetic defect and, also, other diseases are very important. However, this disease is much more prevalent, as everyone has stated, than it is normally assumed to be. And, therefore, we feel funding is extremely important to alleviate the fear of the families who may have this genetic defect, as well as the care of the person who already has it. As was stated that the families...we do have the daughter that comes in to visit the mother and she is very worried as to what might become of her and what might become of her family. And if there were some sort of a clinic, as you stated, to take this daughter to, so that she could talk everything over with everybody--and I know there are no answers to this at this time, but if we did have extra funding we might be able to have different counselling and clinics. Because at times we do spend time with her and talk with her, but we are so busy and we have so much to do basically, too, for the patient, that there is not always time that we can spend with her that she would really require. And also funds would help find a cure or aid in periods of time of remission with the financial burden, because these patients, before they get to this stage, will have times of remission that if, maybe, they did have the fundings, they could have physical therapy and they could have speech; this could possibly help. Now, this patient, at this stage now, there is no way that you could ever use physical therapy or P.T. But, maybe, if there had been money for this patient earlier, she might not be at this stage at this time.

Thank you.

N. WEXLER: Was there anything in your training that would have made it any easier for you to work with either this patient or an HD patient earlier in the disease?

RUTAN: Earlier, yes. As I had stated, that possibly in the training, if they had stated--maybe possibly if we could have given her extra range of motion.

N. WEXLER: I mean, in your own nurses training.

RUTAN: Oh, my own nurses training?

N. WEXLER: Yes.

RUTAN: Well, now, I graduated quite a few years ago--over 20 years ago. And I can safely say that I never saw a case of Huntington's chorea at that time. Now, at this time, that I have been working, I have seen more and more. And, basically, I feel that possibly that these people had--at that time, they had this disease but they were hiding and not, possibly, bringing it out into the open; and not much about it was brought into our training. Neurological diseases were brought in but said that there was no cure and that there was never any P.T. or any physiotherapy brought for these patients at that time.

N. WEXLER: At this time.

RUTAN: So, now, at this time, that's why we feel that--everything is so much more modern and all of these diseases are brought out into the open that training schools now should bring it into much more as one disease.

MCKHANN: Lois Reed, Muriel Wright and Dr. Richards.

I would make one comment and that is that Marjorie Guthrie had to leave temporarily to appear on television at noon and will be back very shortly.

Dr. Wastek, can I ask you to come up now. I gather you have to leave, and I'll ask Lois Reed to wait just a moment.

TESTIMONY OF
GREGORY J. WASTEK, M.D.
UNIVERSITY OF ARIZONA

WASTEK: My name is Greg Wastek and I'm from the University of Arizona in Tucson, and I don't have a prepared statement. I did submit two articles that we've done. I'm involved with basic research in Huntington's disease, and I've submitted two articles, plus a chapter in a book that we're publishing, to the Committee, and hopefully they'll read it later. And the work I'm doing--what I'd like to stress here briefly in my talk, which is totally ad lib--is the fact that we're taking a multi-disciplinary approach in Arizona

We have a clinician, Dr. Larry Stern, who is a neurologist. We have my major professor, Dr. Henry Yamamura, who came from Johns Hopkins where Dr. McKhann did, and started the work that we're doing

with Dr. Solomon Snyder there, and we also have a Department of Physical Therapy that works with us, the entire facilities of a hospital available to us, as well as a basic research unit. And what we're looking at, primarily, are neuro-receptor deficits in Huntington's disease. We're studying the neurochemistry of transmitters, the chemicals that carry information in the brain. We're looking for changes in these transmitters as well as changes in the receptors that these transmitters bind to.

And without going into a lot of boring details, all I'd like to say here is that we have found very specific receptor deficits in Huntington's disease, primarily involving the cholinergic system, the GABAergic system, and it appears now even in the [diltmanergic] system. And these are all neurotransmitters that are involved in the caudate nucleus, or in the basal ganglia, the primary area of the motor disorders in Huntington's disease.

And what I would like to say briefly here is the fact that I would like to stress that we need these types of multidisciplinary centers that we have. We have access to patients while they're in the hospital, so that we can study the clinical manifestations of the disease. Once the patients have expired, at autopsy, we can have immediate access to Huntington's disease tissue which we then use for our neurochemical studies. And when we're through with these studies, we can go back and correlate this with the condition of the patient while they were alive, what medications they were taking; we can correlate these with receptor deficits, neurotransmitter changes. And we can try--which we are trying right now--to find ways to offer a symptomatic relief to Huntington's patients while they are alive.

And I think that we also want to emphasize the fact that we're looking for some type of predictive test in Huntington's disease. And I think we're well on the road toward that now with the GABA-ergic system in particular. We think that we have developed a fairly sensitive assay for cerebro-spinal fluid, for we can detect low GABA levels in patients with Huntington's disease. Now, this is very tenuous, very experimental, but if it does work out, it's obviously an extremely valuable tool. We can tell people which offspring of Huntington's disease patients are at risk for the disease; we can use it for genetic counselling, and we can also use these types of tests to monitor levels of drugs in the brain itself while we're treating people for Huntington's disease. So I think that's about all I have to say, and I just wanted everyone to know the fact that there is a lot of research going on, and the fact that the tissue we get from both Huntington's disease patients and normal patients is very valuable in making comparisons with the normal and the diseased state. And by comparing these two, we're able to, I think, really get at the heart of what the disease is and try to make some advances toward, if not curing it, at least helping the patients while they're alive.

MCKHANN: Dr. Wastek, I've heard the comment made that our knowledge about the basic mechanism in Huntington's disease is about the same state that our knowledge of Parkinson was, say, some 10 or 15 years ago; and that many of us are hopeful on the basis of this, we could see similar advances. Do you agree with that assessment, more or less?

WASTEK: Yes, I would. As a matter of fact, when L-dopa was first tried, as you well know, as a treatment in Parkinson's disease, it was an abysmal failure. But, consequently, through increasing dosages and continued research, it turned into a fairly valuable tool. And I agree with your analysis right now that Huntington's disease is at the point where we're defining what the basic deficits are, and we're trying to come up with certain drugs that will alleviate these deficits. And although certain trials to date have been equivocal, I don't think that's any reason at all to give up on them. And I think that the more we learn about these systems, the more we'll be able to design drugs around the ones we already know and possibly come up with an L-dopa for Huntington's disease.

LI: You talk about your predictive test based on spinal fluid --how early can this test be performed? At what age?

WASTEK: Apparently, there is no correlation with the decrease in GABA, the neurotransmitter, and the duration of illness. This was a surprising finding to us. So, consequently, we feel that we can use this almost immediately in someone who is at risk for Huntington's disease before they've shown any symptoms and while they're still fairly young. But this is just a projection on my viewpoint. I really have no evidence to support this, other than the studies that we've done already and which are going on right now.

LI: When you say "fairly young", you mean, say, under 10 years old?

WASTEK: I would say so, yes. I would say it's a very reasonable prediction.

N. WEXLER: In the center that you describe, do you just have a clinical research center where you bring in patients periodically for research, or is there some place near by you where patients live full time?

WASTEK: Okay. Well, Arizona has primarily two large cities, Phoenix and Tucson; just like Denver is the big city in Colorado. And Dr. Stern has started a specific Huntington's clinic and we have patients coming in from all over Arizona, from the surrounding mining towns and whatnot, and even from some parts of Southern California,

and they come in primarily as inpatients in the hospital. Now, I know he does see a few on an outpatient basis, and I'm not a clinician, so I'm really not qualified to talk about that. But the fact is--the fact of the center approach being so valuable is what was emphasized here before--is the fact that most physicians don't see Huntington's patients to begin with, and once they do see them, there's no followup. Where, here, we can follow someone from when the disease--and possibly with the CSF assays--follow changes in neurotransmitters. And once the person has died, then we can use the tissue to make further studies and correlations.

N. WEXLER: They will come back, let's say, from some small mining town to your center if they're in a terminal stage?

WASTEK: Oh, yes, definitely. I think a large part of that is due to the earnestness of the clinicians in treating these people. They let them know they want to see them; they're interested in the disease; they want to find a cure for it, if possible. If not possible, they want to make these people more comfortable during the course of the disease. And I think that this makes people more than willing to come back and sacrifice the time driving back and forth if they have to do that. As of now, there are no facilities for staying there, other than literally checking into the hospital.

N. WEXLER: How does Dr. Stern manage to do this for free, as I hear he does?

WASTEK: He just enjoys it; he makes time to do it.

N. WEXLER: The hospital there supports him to do this?

WASTEK: Oh, yes, yes. As a matter of fact, I am very impressed by the cooperation between clinicians and basic scientists who many times aren't known to get along too well on research endeavors. But here, there is no problem; there is a willing exchange of ideas and tissues and it's an enjoyable atmosphere to work in. I think a lot of good work has come out of it and will continue to come out of it.

Thank you.

TESTIMONY OF
MURIEL WRIGHT
DENVER, COLORADO

WRIGHT: I'm Muriel Wright. I have a niece who has Huntington's. My brother-in-law had it. He was diagnosed at Colorado General quite a few years ago, and he lived eight years at Pueblo State Hospital and then was transferred for his last two years up to Westminster Nursing Home. My sister never saw her husband in this condition because she divorced him before this happened; and they had six children.

And one of my concerns now is, there are five other children involved, ages 22 to 34. There's a possibility that he was not their father. One of my nieces went to her mother when I was there and she asked if there was a possibility that Kenny was not her biological father, she would like to know because of the Huntington's. And my sister--I mean, no one was questioning her morals. It's just the idea that, you know, these five kids have this question on their minds now that their sister has it.

And I spoke briefly to our doctor yesterday, to see if there were any tests that could be performed to see if he was their father, but he died in 1968. And he said he did not know, you know, since he was already deceased, if there would be any blood tests or that they were more intricate than just normally checking parentage. But, anyway, that's the five, because one is married and he's hesitating about children; the other one isn't; and Marilyn, my niece that is affected, gave birth to five children with her first marriage and this ended in divorce because she was beating the one boy and mistreating, in general, the children, especially this one. And he has to take pills for the rest of his life because of the damage she did to him. And, anyway, before their marriage ended in divorce, the neighbors reported her and she was given a choice between jail and mental hospital; and, of course, she went to the mental hospital and, apparently, was made to realize what she was doing. Apparently, she didn't realize and, anyway, it was too late then to patch things up and the father had custody of the children.

And about a year later she remarried and had a child that was born with a few little difficulties which were straightened out. Her second husband died in 1974. Before the year was up, because of her trying to find a husband and a father for Cindy, who was about three years old, her sister-in-law took the child away because neighbors reported too many cars from men overnight and everything.

So, she is completely alone which is very bad because no one in the family wants her around for obvious reasons--I mean, the move-ments, and so forth, that she goes through. She is by nature a very stubborn person to begin with and so it is difficult to help someone like this now that she has this sort of disease.

I did try to--I mean, to me, it was very difficult--but just a few months after her husband died, I did try to point out to her that there was something wrong with her; that I wanted to visit her. (She lives in California.) I wanted to take her to a neurologist there, or have her come back here and be looked at at Colorado General. And it was one excuse after the other, but I did write to her doctor when I got back and the letter was never answered, although he did tell her that I wrote and she wrote me a few nasty letters; but then, I realized she was ill.

When she did come out here and visited her sister, she was diagnosed, more or less, as a Huntington's. Her sister had her for awhile and then she didn't want her; her mother...she went back to Indio, California, and she was actually put out of her home. The welfare man came, took her to the bank to get her money for the bus, put her on the bus, and ran her out of town, more or less, which I think was unfortunate because she had this paper that Dr. Margaret--somebody over at Colorado General--she gave her a paper stating that she had a neurological disorder, which caused stumbling and speech difficulties, and so forth. Because she was...well, people would think she was either retarded or drunk. And it's been hard for her, because she has no one. She doesn't have a devoted husband--like Mr. Wilson is--since he died.

And I'd like to know if there isn't some way...I've tried to get her to get in contact with the group out in Riverside. There should be some mental health counselling, because she is still looking for a man. If this is...well, she's been beaten up; she's been raped. There is still a possibility of her having children and she could have some sort of counselling and then, you know, some sort of operation so that there would be that. Her mother won't look in on her; it's really a sad situation. I mean, it's bad enough that they have people that they can turn to, but in her case, who does she turn to? There is no--I don't think there is. If I knew how to contact somebody to go contact her, I would love to do this, but I can't go out there and take care of her myself, because I saw her father in his last stages, and there was no communication available and all these things, and it's rather upsetting.

N. WEXLER: I think you just pointed out the need for having some kind of facility where somebody who doesn't need institutional care can still go, if they don't have a family.

WRIGHT: Oh. I'd hate to see her go to a nursing home before she needs it. And when she was out here, her sister was trying to get her power of attorney and put her in a nursing home. But she wasn't ready for it yet; there should be an in-between place.

N. WEXLER: There should be.

MCKHANN: Dr. Richards.

TESTIMONY OF
R. B. RICHARDS, M. D.
FORT MORGAN, COLORADO

RICHARDS: I'm Dr. R. B. Richards; I'm a medical doctor. I've been in general practice in Fort Morgan, Colorado from 1935 to this date.

My experience with Huntington's disease is really very limited. But over a long period of time in my professional practice I've had only one patient who was afflicted with this disease. The time I saw her, she was in a rather advanced age, and her disease was advanced and she shortly died from the mental and physical deterioration. However, my long experience has been with my wife, who recently died from Huntington's disease.

I'm sure what I'm going to say you've heard over and over, but in my experience, it has a very subtle onset of mental and emotional disturbances. These things increased over a number of years and during these periods these reactions became very devastating to her, to myself, and to the children. Her physical disability became manifest at a considerable length of time after the mental part of this. But both, mental and physical, increased in severity making her a total mental and physical cripple. This demanded total nursing home care from 1964 to her death in 1965.

I will say that I felt she had, up in our community, excellent nursing home care. There is no problem; the personnel of the nursing home were very devoted. They seemed to be devoted, particularly, to her in the care that they gave her. Also, at the time of her death, I did donate her remains to the University of Colorado for research at that time.

I also have a son who is a practicing neurologist, who, as far as I can tell, is bravely facing the future. But I really cannot evaluate what his thoughts are regarding this future of his. I definitely feel because of the ravages of this disease are so devastating, that an all-out research of the genetics should be carried out. If such research could lead to the identification of those that would not be afflicted, it would release a tremendous burden from them and they could go forward in their life's work without this shadow over them. For those that are afflicted, I feel that they should not reproduce and that they should have counselling for the development of artificial families, so to speak.

This disease is a tremendous problem, of course, involving genetic and medical, psychiatric and social phases. I feel that it cannot be totally divorced from the other degenerative neurological diseases. I think there could be interrelationships some way, particularly in scientific research. I feel that all of these phases of this disease deserve national attention and research and I think that's about all.

I'll be glad to answer any questions.

MCKHANN: I'd be interested, Dr. Richards, in your assessment of the knowledge of this disease, related disease, among your colleagues.

RICHARDS: I think that because my wife had it, in my small community my colleagues are acquainted that there is such a disease, but, as I said in the beginning, I have only had one patient in my career with this, and the rest of them have had none. And so I'm sure their information is practically totally lacking regarding this.

LI: How many children do you have, and how old are they?

RICHARDS: Oh, I had one adopted daughter; we had one natural son. He is the physician, yes.

MCKHANN: I'd like to ask Lois Reed, Dr. Syner, and the other Dr. Lubs if they would come on up.

Ms. Reed, we'd like to ask you to go first.

TESTIMONY OF
LOIS REED
PRESIDENT, ARIZONA CHAPTER
COMMITTEE TO COMBAT HUNTINGTON'S DISEASE

REED: I am Lois Reed. I'm President of the Arizona Chapter of the Committee to Combat Huntington's Disease, I'm from Phoenix, Arizona.

I want to say first that you probably haven't received as many letters as you might have expected. And I'm amazed at some of those who have testified here, who apparently did find time and energy to write their letters, because I'm sure the ones you should hear from, you haven't heard from. If there's a patient in the family, I'm sure that the family is so overwhelmed by the problems, and they're busy taking care of this patient or running back and forth to an institution to see the patient. And those who should be writing their letters probably haven't gotten them written. We had a meeting--the Committee to Combat Huntington's Disease holds meetings every month. And at our last Friday's meeting, the purpose was to get together with some of those who had found it emotionally impossible to write their letters and we helped them write their letters. We got five letters written, but what I'm going to do is read the letter that I had written to the Commission, because if I try to just talk about it, I probably would be here for two or three days. This might slow me down and stop me.

Briefly, six years ago, when my niece stood beside her father's deathbed and said, "Aunt Lois, we finally found out what Daddy has; it's Huntington's chorea," I asked, "Huntington's what?" I thought it was something he had brought back from Korea. I say this because in my work as CCHD president, I do many, many things. We don't have all that many volunteers. And in one fund-raising event, this

fellow said, "Yes, it sounds tragic, and I'll be glad to donate. But what I want to know is"--he didn't know it was in my husband's family--and he said, "What I want to know is why do the stupid so-and-so's continue to propagate?" So, of course, I told him, "Well, I don't know why the other stupid so-and-so's did. I can only tell you why we did. We didn't know,"

My husband's family has never been very close and we've never lived in the same area. We had gone back to Illinois for his mother's funeral and were vaguely aware of the fact that four of his aunts and uncles had died under disturbingly similar circumstances, most of them after spending years in a mental hospital. One of his sisters had by this time been committed to the state hospital, and a third sibling was showing symptoms similar to the others. However, our doctor had given us a report before our marriage that his mother's problems were--and I'm sure hundreds of you have heard this over and over again--quote, "a nervous condition brought on by the menopause and aggravated by divorce--nothing hereditary." We had six children of our own and five grandchildren before we learned differently.

All the death certificates had listed cause of death as either heart failure or pneumonia. So, after my brother-in-law's death, we contacted--thank God--the National Committee to Combat Huntington's Disease. And we learned more about HD. I had read an article in the paper after we came back from that visit, describing Huntington's chorea--and I'm sure many of you know what it did to me when I saw these words, "100 percent fatal; so hereditary that each child has a 50-50 chance." I went around there like a zombie for a week or two looking at my kids. Let's see, the uncle was red-headed, it'll probably be the redhead that will get it.

Well, during the next two or three months, my youngest son who was 17 threw away everything he had worked for--his scholarships and everything--what's the use?--okay. At that time, none of my kids would let me tell anyone that there was a history of this disease in their father's family. The 17-year-old wouldn't have been able to get a date; the others couldn't have gotten good jobs, et cetera.

So then, I contacted the Committee to Combat Huntington's Disease and I learned that if my husband does not have the gene--which we certainly are beginning to believe he doesn't have--he's 62 years old now--then I learned that it doesn't skip generations; that all of our children are probably free and our grandchildren. So that's why I got into the Committee to Combat Huntington's Disease. I couldn't face my God, or his relatives if I didn't do what I could to help alleviate the suffering of the others,

During the past six years, I have served--naturally, without pay, as all the rest of them do--as secretary, then as executive secretary,

and now as president of the Arizona Chapter, and National CCHD Secretary. During these years, I have become personally acquainted ...and this...I was interested to hear all these doctors say, "I've only seen one patient; I've only seen two patients." And I've been on television with doctors who have turned to me to ask me questions about Huntington's disease, because I have become personally acquainted with perhaps 100 families who are affected by this disease. Daily I receive calls and letters from people and I thought of this when Mr. Wilson spoke. I receive calls and letters each day from people asking for help or advice, and the fact that I'm seldom able to offer very much concrete help or advice is beginning to get to me. I am praying that through the Commission, at least a measure of comfort and assistance for these families may be forthcoming.

And I want to tell you just a little bit about Sandy Lambert. I first met Sandy, who is now 29 years old--I first met her five years ago, soon after her four children had been taken from her and placed in a foster home. Now, Sandy...and she had begun to drop the baby. Nobody knew what was wrong with her; her husband had deserted her. And naturally the thing to do, get the kids out of there, because she wasn't able to take care of them. So, her children were placed in foster homes.

And Sandy is more fortunate than some, having found a fellow who has enough severe problems of his own that he has found it possible to love Sandy and to form a relationship with her, in spite of her very visible illness and her speech difficulties. Sandy isn't now totally alone, as so many are. She has been able to postpone institutionalized care, although the conditions under which she and Joe are living leave a whole lot to be desired.

And if only there had been some kind of home health care available to Sandy, she might at least have died with her children nearby. She doesn't even know where three of her children are today; she does know where one of them is. I see Sandy weekly.

Now, I want to...I made a little note here of something I thought of...it should come at the end of the next session, but I'm going to say it now. On this next patient--I had been so concerned about her so many times, and I finally located a doctor who had cared for her in the state hospital and I said, "What can I do to help this lady?" I didn't know much about Huntington's disease at that time. All I got from him every time I called was, "Do you want a petition to commit?" No, I didn't want a petition to commit.

Paula...and I saw her at this Friday night meeting...we wrote her letter. And she said, "Certainly, use my name." Paula Bryan was in a boarding home where she had been placed by Arizona State Hospital authorities when I met her. Paula was employed as a domestic in a doctor's home at the time of her diagnosis. A friend, trying to be helpful, informed her employer of the nature of Paula's

illness. The employer immediately let her go and Paula registered in a motel under an assumed name and took some pills. She was discovered and taken to a Phoenix hospital where her life was saved and she lay in a coma for several days, unidentified until her former employer's husband, a doctor, happened to view the comatose patient and recognized her as the former employee in his home. She was ultimately committed to State Hospital, later released in the company of a gentleman who himself had been an inmate there. They lived together for a year or so, until the fellow assaulted and beat her severely. Since that time Paula has lived alone in unbelievably horrible apartments, trailers, which are all she can afford on her income. She gave up her only luxury, coffee--she loved coffee. Several months ago she gave up coffee because there wasn't enough money to buy it.

Some of us...we do not give direct financial aid to the Committee to Combat Huntington's Disease, but most of our board members and many of us sneak by and leave her a pound of coffee now and then out of our own kitchens. She did try suicide again last year, but she was again denied relief from her suffering; they found her too soon. She's 40 years old now and she realizes that her disease is progressing rapidly. She's fighting against any kind of help which she feels might hasten the time of her commitment to an institution. She watched her mother spend 13 years in an institution, abandoned by all the members of the family, except herself.

I don't know why...yes, it would take too long to go into why families do abandon patients. And she is unable to stay alone, but she is, as I said here, fighting against any kind of help which she feels might hasten the time of her commitment to an institution. She falls frequently; she's been picked up by the police on several occasions and is, of course, in danger of being injured while trying to do her own cooking and housework.

Lila Gangi, a once very, very beautiful woman, who was runner-up in a Miss Arizona contest a number of years back, was sitting in a chair at Barrow Neurological Institute in Phoenix eating lunch when I first met her, getting food in her face and her hair and on her lap because of the chorea movement. Her speech was almost unintelligible even then. Her husband, finding it impossible on a mediocre salary to hire help in the home for her care and for care of the three children, had his wife committed to Arizona State Hospital with the understanding that a temporary commitment was the way to force his county into giving financial aid. He was later unable to get her released and called me on several occasions, feeling helpless when he went to visit his wife, who, he told me, was confined in an area with people of both sexes, in various states of dementia, who, among other acts, were exposing their bodies and in many ways adding to the emotional suffering of his sensitive wife, who I am convinced had very little mental disturbance other than the speech difficulty.

Lila was ultimately placed in one of the State Hospital boarding homes. Her husband, overwhelmed by the financial and emotional burdens and the feeling of frustration and helplessness each time he visited his wife, finally gave up the infrequent 500-mile trips to visit his wife. Lila died alone at the boarding house last summer. In this case, also, the right kind of care in the home would have allowed her to have at least died surrounded by her loved ones.

Now the Committee to Combat Huntington's Disease has accomplished a lot in the area of education, but there is still much to be done. Very few who care for Huntington's disease patients really understand the disease, and the general public in many cases still thinks that HD is another name for Parkinson's disease.

A woman who lost her husband to HD recently went to her family doctor. I know this woman well. This family doctor had cared for the Huntington's disease patient for 12 years prior to his death. After his death, a year or two later, Rachel had occasion to go to her doctor for neuritis or something, and he said, "How have you been, Rachel?" And she said, "Oh, I'm so upset. I know that Donna is now developing symptoms of the disease." He said, "My dear Rachel, you have suffered so much and this is one worry you need not suffer. Didn't you know that only males get the disease?" It's hard to believe that our medical profession is still so uninformed.

I feel strongly, of course, that continued and better funding of research is terribly important. But in my opinion, at this stage, after spending six years working with these families, I think that right now we have to concentrate on emotional and financial help, for not only the HD patients and their families, but those who are at risk. And I'm sure I've demonstrated the need for education of the general public and the health professionals.

Last week I was in Tucson, Arizona for a meeting and while there, I always try to get on the phone and call a few of the Huntington's families that I have met previously...and he also gave me permission now--three years ago he wouldn't--to use his name--none of you know him anyway--Reverend Jack Kelly, whose wife died six months ago of Huntington's disease. And in this conversation, he acted like he was making some terrible deep, dark, black confession. He said, "I don't know that you know this, but I found it necessary--we found it necessary a year or two ago to dissolve our marriage." This is a minister of the gospel telling this--because, he said, "I was totally unable to pay \$900 a month that it was costing to care for her in this home."

And I, right now, know of seven families who have found that they were forced to get a divorce in order that the sick spouse might have the care that they needed. I had a family call me two weeks ago and say, "Lois, I want your advice." I said, "I can't

give my advice on that." They had talked it over. He was just diagnosed a few months ago. He's fine, but they have already decided...they didn't want my advice; they wanted my approval, I think...that they would get a divorce immediately in case he had mental problems. They would do it now so that she wouldn't be saddled with the financial responsibility of caring for her husband when he got bad.

I'm going to quit; otherwise, we'll be here for weeks and weeks and I'll keep talking.

MCKHANN: I have some notes here, but I gather one of the things you were coming down pretty hard on is something that Mrs. Wright had mentioned to us too; and that is, the need for home care support. In other words, an alternative to institutions.

REED: That's right. I feel that...may I enlarge on something I said? In many cases, I think, in most cases, I believe the patient is better off at home. But in so many cases...this same Rachel Wilson I mentioned to you, breaks into tears and she said, "Yes, you're making me feel terrible. I had him placed in an institution," and she burst into tears. She said, "If I had only had help, even two days a week, I could have kept him at home."

And I want to make another observation, because I probably...I don't want to sound egotistical or something, but I know I've seen more Huntington's patients than most doctors and visited in their homes. And I am convinced that there's not as much mental problems as a lot of people think. In many cases, I think that the mental problems have been caused by the situations and the circumstances under which the patient has to live and the problems of speech communication and that a lot of times they assume that there is more mental disturbance than there is because of the speech difficulty.

N. WEXLER: Lois, in all of your six years of magnificent work, have you ever found a family that has had home help, or that has had really good graded facility care?

REED: No, I have found families who were very fortunate in that they were financially able to hire help in the home. I have not.

N. WEXLER: At a middle-income level, you haven't had anyone that's had that kind of help?

REED: No way.

N. WEXLER: And this is a lady with lots of experience.

MCKHANN: Dr. Syner?

TESTIMONY OF
JIM SYNER, M.D.
MEDICAL CONSULTANT
COLORADO MEDICAID PROGRAM

SYNER: My name is Jim Syner. I serve as a medical consultant with the Colorado Medicaid Program and I was asked to outline the medical services that would be available through the program to any patient afflicted with the disease.

Having sat in on a fair proportion of the conversations this morning, I would recommend to the Commission that in other areas it might be well to request that one, a psychiatrist who serves as a consultant to a State Medicaid Program might be a more appropriate selection than myself as an internist, because in the specialization of work with a program as broad in nature and scope as Medicaid, there is great emphasis on specialization and a consultant who serves for psychiatric illness might have more direct knowledge and experience and exposure than I, as an internist, would. I just offer that as a suggestion.

But just, first, let me briefly say that for those who may not know what the Medicaid Program is, just let me take a moment to at least give you that much orientation. The Medicaid Program is a medical program which is sponsored by both the Federal and state governments. It isn't exactly the equivalent of Medicare sponsored by the Social Security Administration, so that there are great differences between Medicaid and Medicare Programs. The single greatest difference is, of course, the population of individuals served by that program. Generally speaking, the Medicaid Program serves the welfare recipients. There are exceptions to that in terms of certain criteria for medical eligibility for the benefits, but, generally speaking, that will give you an orientation as to who are the individuals that are eligible recipients of the program benefits. One other point about the Medicaid Program, it is administered under the provisions of a specific law of the Social Security Act known as Title XIX.

The medical benefits that are available to any patient who has this disease under the program would cover any of the services for medical care that we all know and understand. Number one, it provides for physician services, whether that physician service be provided in the doctor's office, in a clinic, in a hospital, in a special center or in a nursing home. The program provides for inpatient hospital care, and the benefits in inpatient hospital care would be those that are medically necessary; those that are considered to be required as seen by the patient's attendant physician; then

outpatient hospital care. Nursing home care is a benefit of the program; laboratory, X-ray examinations are benefits of the program; certain kinds of medical equipment, respirators, wheelchairs, et cetera, are benefits of the program; transportation to and from the doctor's office, to and from a facility for medical reasons, are benefits of the program. There is a benefit of home health care under the Medicaid Program.

And I do want to just make some qualifying remarks about that, because I've heard statements here today that lead me to believe that this is an extremely important issue and an extremely important concern as far as the care of these individuals are concerned. There probably is no area of the Medicaid Program that is more in need of a lot of careful thought and a lot of careful planning than the home health care component of the Program. This is due to a multiplicity of reasons, and I think one of the key reasons that I've heard here today is the fact that so much of home health care is not delivered or provided by a doctor. It's provided by either members of the family, home health attendants, or other kinds of providers of services.

The way many of the rules and regulations governing the program are currently stated, it is difficult to facilitate the full battery of services that are required in the home, because the program...so much of the program is oriented to hospital care, inpatient hospital care, outpatient hospital care, and direct physician care. Whereas, more and more, we're learning that there is a tremendous need to involve other kinds of services, particularly in the home setting, and the people who provide these services have to have an independence, if you will, and they have to have an expansion of their role that permits them to do this. In other words, if you delegate the responsibility to do something, you have to give them the authority to do it. And by and large, within the context of the Medicaid Program, so much of the authority to do things is tied to licensed physicians. For instance, we cannot utilize a registered nurse as a direct independent provider in a home, and there's other kinds of people we can't. So that's a major issue that goes back to the structure of the law itself from which the rules and regulations emanate.

Now, recently, the Federal government did amend the regulations governing home health care, at least for the Medicaid Program. And as I read those regulations, I see tremendous opportunities to be more flexible and to be more innovative. There's still administrative and financial problems that have to be faced to get those regulations properly implemented, but at least I do feel optimistic that at long last we have some opportunities to establish much more meaningful programs of home health care.

N. WEXLER: Oh, could I ask one question? Could you, perhaps, give us any recommendations as to how we might have input in that process? If the regulations are changed sufficiently, is it a matter of suggesting certain specific programs of home health care?

SYNER: I think--to me, at least--there is a major technical problem in Medicaid; and that is, each and every individual state has different statutes that govern the delivery of services. But I do believe that if there was closer liaison between representatives from the Federal government, DHEW, and states, particularly as it relates to the several regulatory boards that govern what one can and cannot do, that this would be tremendously important in beginning to get the thaw, even if it was just applicable to the Medicaid Program. But, to me, that's the big technical problem that you run into, is anytime you cross a state line, they have their own individual statutory regulatory requirements that govern the practice of medicine or the other people that come under regulatory jurisdictions in the state. That's the problem. I think that at the Federal level an enormous effort has been made to provide independence and expanded roles to a large variety of health care providers, but we run into that problem at the state level.

N. WEXLER: So that if our Commission were to make a recommendation, for example, we would need to make it at the Federal and at the state levels?

SYNER: Yes. Arizona is the state that doesn't have Medicaid.

N. WEXLER: Yes, I'm aware of that. If you have any other specific suggestions as to how we can make sure that our recommendations get down to the state levels, we would appreciate very much all specifics.

SYNER: From the point of view of having an organizational nucleus around which you can move, I would strongly recommend that you go directly to the Director of the Medicaid Program in the state. Let that serve as an organizational structure, now, to get to all of the component parts that you have to get to and kind of build your program around that approach. Because, certainly, we--say, in Medicaid in Colorado--have the variety of linkages that would be required to get into the structure. At least for openers, that would be my suggestion.

N. WEXLER: One difficulty that we've been hearing repeatedly is that even though Medicaid probably provides more services than any other Federal program, that the eligibility requirements are so steep that you have to be totally indigent to qualify for Medicaid services. In a family with the genetic disease, it makes it almost impossible to save money for future jeopardized generations.

SYNER: Well, you know, you might consider the designation of people with Huntington's disease as categorically needy. You know, we...well, aid to the needy blind, aid to the needy disabled--I mean, that is a recognized precedent. And if at the Federal level they expanded the definition of categorically needy to include this group,

then you would have accomplished what you want to and--you know, we're constantly playing with definitions to meet needs. We have to. So you expand the definition.

For instance, a hemophiliac in a way becomes...we have several individuals in Colorado who have hemophilia...and at certain levels of disability they become categorically needy. So at least it's something that you might consider and work with people who deal with that kind of technicality of language, because the intent is to make the benefits available to them. So, just get the language that implements the intent.

N. WEXLER: That's an excellent suggestion. We'll look into that. Thank you.

TESTIMONY OF
MARIE LOUISE LUBS, PH.D.
ASSISTANT PROFESSOR OF PEDIATRICS
UNIVERSITY OF COLORADO

LUBS: My name is Marie Louise Lubs, and I'm an Assistant Professor of Pediatrics at the University of Colorado Medical School. I am particularly interested in genetic counselling, but I'm here today to say a few words about psychological aspects of counselling in families with Huntington's disease.

Genetic counselling generally means giving genetic and clinical information to families with inherited disorders. Genetic counsellors usually have no special training in clinical psychology, but are generally aware of and sensitive to the psychological aspects of the burden of genetic disorders. I can think of no other --no genetic disease where the psychological burden, as well as the genetic risk, is greater than in Huntington's disease. Although I would like to see a psychological approach applied more often in many genetic disorders, a very good place to start, therefore, is in families with Huntington's disease.

By the time an individual at risk sees a genetic counsellor, he or she is generally very familiar with the clinical prognosis of the disease. The genetic risk in numerical terms is also not very difficult to convey. But to leave the individual, once he has understood this factual information, is clearly to deem him "cold turkey." One thing that makes the disease particularly difficult from an emotional standpoint, aside from the clinical symptoms of which you have heard previously, is the variable age of onset of the disease. The age of onset of the symptoms varies between about age 20 to about 70. Half of all individuals with the gene show symptoms by age 40. It is noteworthy that the age of onset is generally not familial. In other words, the fact that a parent did not develop the disorder until the age of 65 does not mean that the child might

not show symptoms at the age of 30. There is generally no pattern within a family. Therefore, it is impossible to realistically plan your life or your family around the disease.

Many individuals choose to take a chance and have children, live as normal a life as possible, and hope that they would have escaped the dreaded gene. In other instances, the term "normal life" is totally unrealistic, and the problem of coping with the burden a major task. Even if we, a few years from now, will be able to predict with the aid of laboratory or other tests who in a sibship at risk will ultimately develop Huntington's disease, there will be those who would not want to know, while, let's say, the age of 15 or 20, that they will likely be affected by the disease 20 to 40 years later. Others would, no doubt, be optimists and take the chance of 50 percent of getting a clean bill of health.

But even if we learn how to diagnose a disease before symptoms are apparent, we're still going to have the psychological problems in the individuals at risk. If they cannot function as productive citizens in the society, the burden will also, secondarily, affect us all. I'm sure that it would be economically beneficial not to speak of the individual relief if we could help these families and the individuals at risk to function and live in our society as productive and psychologically adjusted men and women.

I would like to see psychological counselling offered individually, as well as in groups, over longer periods of time--maybe six months, a year, or even more. Since, particularly, group counselling has not been tried before, we plan to apply to NIH for Federal funds to implement and evaluate such a program. I see these groups not consisting of members of one family, but maybe individuals at risk of the same age; maybe even with several genetic disorders represented as well. If we can show long-term counselling is beneficial for the families in reducing anxiety and guilt and improving self-esteem--and I personally don't have any doubts that it would be--we would like to see counselling groups developed in other cities in the United States.

Since we started planning this project, we've asked members of five different families if they were interested in participating in such a program and have received universally positive responses. I might mention that this approach is already being taken in Holland and felt to be very helpful in the coping process.

I would also briefly like to say a few words about privacy and about the wish many individuals have not to be identified as associated with Huntington's disease, or at risk of becoming afflicted with the disease. It is very unfortunate both for these individuals as well as for medical research that the disorder is perceived as

stigmatizing. Partly, this is due to lack of knowledge among the general public. This can only be remedied by better education. The Huntington's Chorea Foundation and the Committee to Combat Huntington's Disease, and maybe Mrs. Guthrie, in particular, are working on educating us all, and hopefully we will soon have a curriculum in human genetics in our high schools. This is another area where other genetic diseases are in the same boat as Huntington's disease. But yet, partly, I'm sure, it's also due to economic considerations. If you were to admit that you are at risk for a serious chronic disease, you might have difficulty getting medical insurance, or worse, be discriminated against in employment and other areas. This can best be remedied by National Health Insurance, which I hope will be available in a not-so-distant future.

In the meantime, I would like to plead for the families, if not to step out of anonymity to the general public, so at least to make yourself available for medical studies. This is the best way that we have to solve the problem and possibly find a cure for Huntington's disease. Even if we ask you to have tests that you might think irrelevant to the disease, they may, in fact, in time, turn out to have been totally relevant. Please be patient with us and let us study you.

Thank you.

N. WEXLER: Where in NIH did you apply for funds?

LUBS: Well, we are in the process of writing up this project right now. I have, in the meantime, done a similar proposal that is pending at Maternal and Child Health Care, dealing with muscular dystrophy and hemophilia, two diseases that I'm a little bit more familiar with and have been working with previously.

N. WEXLER: Because, I think, the kind of proposal you're suggesting is one that has had trouble getting funded through the NIH, and we will be interested in following along.

LUBS: I have been funded for five years now doing counselling research, if you so wish, by Maternal and Child Health.

N. WEXLER: Also, if there were to be a predictive test for HD, is there anything that you would see, as a counsellor, that you would need in order to be able to really work with families and give this test? We've heard one figure that the suicide rate among HD patients and those at risk is approximately $7\frac{1}{2}$ times the national average. That probably is an underestimate. If we had a predictive test available, that figure is likely to skyrocket unless we have some kinds of additional supports. Is there anything that you would see that you would require in order to do your job?

LUBS: I think that it would be more difficult to do psychological counselling among people where we're fairly certain that they're going to develop the disease, naturally. I can see that there are many people who would not like to take the test. So I don't know to what extent a test would change the project, as I see it now. I'm not really quite sure I understand your question.

N. WEXLER: Let me put it in another way. Let's say if this project is not ongoing, not funded--just today--that the predictive test was developed tomorrow, do you feel that you have the resources available to you right now to be able to handle the problems that would come up in Huntington's families?

LUBS: At the present time, we have nothing--we have nothing at all. And if you were to create groups, which I think is a reasonable way to do to help people without exorbitant costs...I mean, personal psychiatrists might do better, but I think group counselling in just talking with other people that have problems of similar nature would help and would not be all that expensive. But, at the present time, there is no funding for psychological counselling in the state.

N. WEXLER: How much is the cost for sociogenetic counselling at your clinic?

LUBS: Well, I heard the figure of \$40 mentioned today...I have been working solely in research and I have, therefore, been giving my counselling for free. But I did check with the clinical psychologist and they charge \$35 a session. Group counselling is, generally, somewhat different and I don't know really what the cost would be. But if one...first of all, I think we should set it up on a research basis and really find out what we can do. If we really do help, or maybe we create more problems than were there before--I don't think we would. And if it shows that we really can help a lot, I think that maybe it would be easier for us to get funds for service in the future.

N. WEXLER: I should mention that most psychologists are not covered by insurance.

MCKHANN: Okay. I think we'll stop now for a bit. We are a little bit behind schedule, not seriously so, but Dr. Li and I have plane problems trying to get back to the East Coast. So that we'll, I think, only break for about half an hour.

(Recess.)

MCKHANN: Three names that I have are Ms. Lightbody, Carol Winkler, and Rosalie Dugan.

TESTIMONY OF
MARY LIGHTBODY, R.N.
VETERANS ADMINISTRATION HOSPITAL
FORT LYON, COLORADO

LIGHTBODY: Good afternoon. I'm Mary Lightbody; I'm an adult nurse practitioner at the VA Hospital in Fort Lyon. And for those of you who are not familiar with Fort Lyon, it's approximately a 550-bed neuropsychiatric hospital. We're a long-term hospital and we care, primarily, for the chronic neuropsychiatric male patient.

At the present time, we are home to 14 patients, men age 31 to 63, with a primary diagnosis of Huntington's chorea. Four of these men are in the late stages of their disease and are housed on an acute medical facility. All of these men have had multiple recurring episodes of infection. Four other men are no longer ambulatory, have had no acute illness and reside on 5-B. Five-B is a medical ward and is relatively sheltered. It provides close observation and supervision, plus the necessary aids for feeding, bowel and bladder care, skin care, and offers a limited amount of physical therapy. Six men reside on 4-B. This is a combined medical and psychiatric ward where they are still ambulatory, but require less supervision. Most of their outbursts are controlled with medication.

All of the men with Huntington's disease at Fort Lyon have one or more of the following problems which led to a diagnosis: Choreic or athenoid movements; ataxia; [dysarthria]; defective memory, judgment or orientation; mood defects, such as depression; assaultive outbursts; and emotional lability. Many have one or more of the aforementioned problems with a positive family history. Each of our men has two or more relatives with the disease. In one instance, four generations are involved and this is recorded. Six of these men have no children; the other eight have one or more. The largest number of children in one family is six. Of that particular family, three are already known to have the disease. The average number of children for each of these men is two.

Suicide has been attempted at least once by six of these men, and often on multiple occasions. All have been abusive, hostile, or violent to someone in the immediate family. Length of hospitalization has ranged from eight to 24 years. The average length of stay is 12 years. Some interesting figures--the average cost for 12 years at the present time in our hospital is \$328,500 for one patient.

MCKHANN: That's the total cost effective for 12 years?

LIGHTBODY: Right, that's an approximate cost of \$75 a day. The problems faced by these men are many. Their families are also faced with tremendous decisions and incur the anger of patients and

often relatives with regard to the decision of hospitalizing their son, husband, or father.

Let me list for you some of the problems I have seen firsthand. The depression--most of these men know their diagnosis and the prognosis. They are either aware or past the stage of awareness that their lives will unfold and end in an institution. The frustration--imagine, if you will, being unable to make even a simple movement like putting a spoon to your mouth, and never being quite sure that you'll succeed; of being unable to dress yourself or perform a simple task, such as brushing your teeth. Speech difficulties--the inability to express even simple ideas without repetition, and often the interpretation on the part of the receiver. Anger--severe and uncontrollable outbursts of anger, often with little or no provocation, and often reciprocated by another angry patient. Loss of equilibrium--this results in accusations of drunkenness; falls are imminent, fractures frequently result. Dysphagia and choking--foods often are pureed and having to be fed by another person follows. When the ability to swallow leads to choking, a nasogastric tube is placed--and eventually a gastrostomy tube--a tube into the stomach is the only method of providing nourishment. Imagine never tasting food. Bowel and bladder incontinence--the inability to control bowel and bladder is a tremendous problem, requiring the use of external catheters, medical or mechanical evacuation of the bowels. Muscle wasting--most of the ingested calories are expended on the continuous, often violent movement of the patient. Almost without exception, these men are thin, bordering on cachectic. Large amounts of calories and proteins are necessary. Often the enormous amount of food is not tolerated when ingested. Ulcers or decubiti are a frequent problem, and often extremely difficult to treat for many reasons. As just stated, providing nourishment is a real problem. The disease eventually requires total bed care; turning and positioning are extremely difficult due to the choreic movements. Recurrent infection most frequently seen in our hospital are the URI (upper respiratory infection), or a pneumonia infection. These frequently follow choking episodes and aspiration. Long-term hospitalization in itself often decreases one's natural resistance to this type of illness. Urinary tract infections are prevalent due to long-term bed care and often simple dehydration. Their stomachs cannot tolerate both the food and the fluid in sufficient quantity to meet the unusual needs.

Families that I have contact with have suffered in many ways. They have guilt. "I tried to" or "I should have" are very frequent phrases used. Families frequently fear the patient. They, or a child, have been brutalized. Some children do not understand why the parent was locked away. They do not always remember things the way they were; the beatings, the trip to jail to pick up the drunk; the broken furniture, and the loss of income. Several of the children

I have spoken with are sick with anxiety not knowing whether they will or will not develop the disease. Finances are a real problem. Insurance companies often do not insure known Huntington's disease carriers, whether manifest or latent. Often the disease strikes in the honeymoon or organizational period of a marriage and family. Even a reasonable facsimile of security was never attained due to the early manifestation of the disease. Wives are thrust into the breadwinner status, in addition to their other responsibilities. Finally, the amount of misinformation is almost unbelievable. Some of the statements I have heard are: "Only males are affected." "He got it, now all of his children will have it." "Huntington's skips a generation, so the kids will be okay." Families must be provided with facts. Consequences should be known before the consequences have arisen.

I hope I have given you some idea of the problems faced by these men. At the present time, we have facts and we know the consequences, and this information must be made available. Thank you.

GUTHRIE: Mary, for your information, and others who are here, we have just completed a survey of the VA. There are 500 known diagnosed HD patients in the VA system, and approximately 500 who have been discharged to homes or nursing homes, and who are getting outpatient care. So, you can multiply that figure quite a bit. Can I ask you, do you do any outpatient treatment or care?

LIGHTBODY: I work primarily on an acute medical facility.

GUTHRIE: Do you know if your hospital does do any outpatient care?

LIGHTBODY: We do have three patients with Huntington's disease that are in contract nursing homes at the present time, and that's through our outpatient.

GUTHRIE: Can you describe to us what other therapy you give, other than just medication?

LIGHTBODY: We have physical therapy, which is somewhat limited. Our physical therapy department is very small. We try to keep the men as active as possible with trip-type things--to the canteen, fishing trips, rather sheltered activities, but at least out in the community.

GUTHRIE: Do the Huntington's patients live together with the other neurologically impaired people?

LIGHTBODY: Yes. They are all either completely medical ward or a combination of medical and neuropsychiatric.

GUTHRIE: Do you think that is better for the patient or not better?

LIGHTBODY: Quite honestly, I'm not sure. Because of some of their behaviors, I'm not so sure that it is a good idea. As I said, their sometimes unprovoked outbursts are reciprocated by other neuro or other psychiatric patients.

GUTHRIE: And you realize, of course, that most women or children would not be in the VA system, so we're talking about a lot of people who are not getting care.

MCKHANN: Do you know if the family brings a patient to your hospital, is it difficult to get that patient admitted? Maybe you are the wrong person to ask--you're here,

LIGHTBODY: Well, we only have 14 men, and I would guess that if they had been a veteran, there would be no problem as far as admittance. Some of our men have a very short period of service time and they've been admitted.

GUTHRIE: I have to just repeat that our experience is that some of the VA's do not take the patients in and we have great difficulty. So, if they get into yours, we're very pleased.

N. WEXLER: Do they stay in once they get in?

LIGHTBODY: No. We've just discharged one man to a contract nursing home that I was actively involved in--his planning. And I expect that he'll be out for some time. He has a very interested family. We were very lucky to get a very excellent nursing home for him. And I think he'll get along for quite some time in the community.

N. WEXLER: Do you think it would be helpful if there was some kind of a--either HD or neurological disorders program within the VA system that might have certain kinds of special services for these patients?

LIGHTBODY: Yes, I do.

N. WEXLER: What kinds of services would you want to see in that program?

LIGHTBODY: Well, the veterans themselves need help in understanding this disease and being able to prepare themselves for the outcome. And my particular interest at this point in time is the family.

N. WEXLER: You may have mentioned this before, but is there genetic counselling provided to the family?

LIGHTBODY: No.

N. WEXLER: Is there referral for counselling?

LIGHTBODY: No, not as yet. We're working quite closely with Dr. Armstrong and hope to see that happening.

GUTHRIE: Have you seen our handbook for health professionals?

LIGHTBODY: No.

GUTHRIE: You have not? All right, I would like to make it available to you. We do have a handbook for the care of patients and I'd like you to have it and perhaps let others know about it.

LIGHTBODY: Thank you.

TESTIMONY OF
CAROL WINKLER
EXECUTIVE DIRECTOR
COMMUNITY HOMEMAKER SERVICES, INC.

WINKLER: Good afternoon. I'm Mrs. Carol Winkler, Executive Director of Community Homemaker Services, a voluntary non-profit agency here in the Denver Metropolitan Area which has been providing homemaker home health aid service since 1964. We are an affiliate of Mile High United Way and a third-party contractor for four area visiting nurse services to provide the home health aid service under Medicare. We also receive Title III funds from the Older Americans Act through the Denver Regional Council of Governments, with additional support from private pay patients on ability to pay. We are an approved agency of the National Council for Homemaker Home Health Aid Services. In 1976, Community Homemaker Service provided over 32,000 hours of service to 1,630 adult and children patients.

I wish to speak on the home care as an alternative to institutional care for Huntington's disease patients. Many times during my 13 years as director of the agency I have seen how a well-organized home health team can deliver an array of supportive service to allow a patient to leave a hospital or institution earlier and be maintained at home. I believe we are all aware that patients do better if they can be kept at home in familiar surroundings. I am here to emphasize that the homemaker home health aid is one of the most important members of this health team.

The homemaker home health aid is a paraprofessional with broad training for work with ill, disabled, handicapped people of all ages.

Her--and sometimes his--skills include nurses' aide training in order to assist with the various kinds of baths, shampoo and hair care, skin care, shaving, and assisting the patient to dress. They have knowledge of transfer and ambulation and may assist with range-of-motion exercise when demonstrated by the therapist or nurse. They observe the condition of the patient and report the changes, physical, mental or emotional, to the health team. The homemaker part of their training includes knowledge of good nutrition and special diets, how to prepare the food, how to make it interesting and attractive for the person with the poor appetite, feeding the patient, working with people that have the kinds of problems that the Huntington's disease people do. They are trained in grocery shopping, and how to stretch the food budget. They know methods of organizing their work to accomplish a good deal in a short time, especially in the light housecleaning which they do. The homemaker health aide also changes the bed linen, does laundry, and at times may accompany the patient to medical appointments.

Their training includes understanding the various economic and ethnic factors that influence their work in the home, understanding the effects of physical and mental illnesses, and how to cope with it. They are professionally supervised and receive ongoing in-service training. All of these knowledges and skills give the homemaker home health aide the ability to do her part of the home care for the comfort and recuperation of the patient. The services provided in an individual case are tailored to the need of that particular patient. This is why the homemaker home health aide care can allow the Huntington's disease patient to be cared for at home.

The care to be given by the homemaker home health aide is established in the plan of care which would include doing the activities for the patient that he cannot do for himself, or that he has no one else to do. Over a period of time, the duties are changed as the condition of the patient changes. In as little as a two-hour visit, the patient may be bathed, the bed linen changed, the bathroom cleaned, food prepared. It had never ceased to amaze me how much my staff can accomplish in a very short period of time, and without hurrying the patient.

Their visits also have a mental stimulus for the patient and family. Visits may be scheduled as often as daily if it is needed. Home care cannot replace institutional care, but it is an important part of the health delivery system. It is cost effective for part-time intermittent care. It is not cost effective if the patient is in the need of 8- to 24-hour daily care on a long-term basis.

Community Homemaker Service has provided service to only two Huntington's disease patients. However, I am sure that there are others in our community and across the country who could benefit

from this service of homemaker home health aides. This service is available in many areas of the country.

There are two recommendations that I wish to make. First is that the health insurance companies and the Federal government be encouraged to include for payment the homemaker home health aide service as part of the insurance package, And secondly, that the patients be made aware of the availability of homemaker home health aide agencies and services across the country.

Thank you.

GUTHRIE: I don't have a question. I just want to compliment you on your approach to the problem. I believe, personally, sincerely, that many more patients could stay at home much longer and live better lives and function at a higher level if they had this kind of support very early in the disorder.

MCKHANN: What are the limiting factors in terms of making your services available to more people now?

WINKLER: Money.

MCKHANN: What is your source of money now?

WINKLER: Our source of funding comes from Medicare through our third-party contracts with the visiting nurse service. As a Mile High United Way agency in our own local area, we have funds that we use to subsidize the patient where they pay a fee based on ability to pay. We provide some service through the Title III of the Older Americans Act; and we provide some service through Kaiser [Permanetti] insurance, no-fault insurance. But the dollars are always limited.

N. WEXLER: Can any family apply for your services?

WINKLER: Yes, with one exception. At the present time, we do not use our United Way funds to subsidize service for those that are on Welfare, Public Assistance. In the Denver Metropolitan Area, the counties have homemaker service for recipients. We were organized to provide service to those primarily on low income that had no other place to turn and that we could subsidize the service. If the service comes to us or the request comes to us from a third-party contractor, such as Medicare, Kaiser, something of that sort, then we do provide the service to those on public assistance.

N. WEXLER: Do you have any idea why there are so few HD families that have utilized your service?

WINKLER: I would say probably the lack of information about the agency; as money is always the problem, we do not do a great deal

of publicity. We try to interpret to appropriate groups. If we do advertise, so to speak, our limited staff cannot handle the calls; and I would say it's literally a lack of information.

N. WEXLER: How does a person find you?

WINKLER: As far as a general answer to that question, if you're referring to, say, across the country kind of thing, or specifically here in Denver?

N. WEXLER: Well, both.

WINKLER: Both? Well, specifically, here in Denver, we are listed under "Community Homemaker Service." There is a second listing under "Homemaker" that refers you back to "Community" listing. It is listed in the Yellow Pages under Social Services. If you are in an area wondering if there is such a thing as the Homemaker Home Health Aide Service available, the best contacts are through the Public Health Department or the area visiting nurse services, or the Social Services Departments.

TESTIMONY OF
ROSALIE DUGAN, R.N.
LaSALLE, COLORADO

DUGAN: I'm Rosalie Dugan. I'm a registered nurse. For the past 25 years, I've worked and I have taken care of one Huntington's diagnosed. I'm sure that through the years, we've probably taken care of many that have not been diagnosed.

The other contact I have is a friend who has Huntington's in the family, and I know I've chatted with her many times and it's a very ...a thing that we need more counselling on, more group therapy of people who can get together and have rap sessions to get their frustrations out in the open.

At the time that I took care of this gentleman with Huntington's, we really tried to dig up information, printed matter and places where his wife could turn for counselling. And really, we came up against a stone wall. There really just wasn't this much. Our neurologist who was handling the case really couldn't do much as far as education was concerned.

And I'm all for...my suggestion is that we need more education, both professional and layman, and groups--therapy-type thing--where people can rap out their frustrations; and research for early diagnosis.

MCKHANN: How large a community is LaSalle?

DUGAN: Well, LaSalle is not very big but we're a suburb of Greeley.

MCKHANN: Within that community do you think that the fact that there were few patients with this problem led to a greater awareness at all?

DUGAN: You mean for the public?

MCKHANN: Or the people who are involved professionally--are there people in nursing?

DUGAN: I really--I don't think so at this time; just the ones who are directly involved or have had direct contact with them.

LIGHTBODY: May I make a comment? We were doing in-service for some of the nurses at Fort Lyon on specific neurologic diseases. We put out a Medline search for information on Huntington's disease--and Medline search, for those of you who are not familiar, is a very large library housing all of the--most of the medical information available. We came up with 54 sources; that's a pretty limited amount of information.

N. WEXLER: That was on nursing care on HD?

LIGHTBODY: Right.

N. WEXLER: Is there anything...really, this is a question for the three of you...that you--anything as far as changes in nursing curriculum--that you would recommend that would enable you to care for HD patients better?

DUGAN: Of course, in my curriculum there was nothing. It was a few years back...mentioned, but this was all.

GUTHRIE: So, it should be then in the nurses' curriculum, in terms of study?

DUGAN: I think it should be, yes.

GUTHRIE: I'm just curious...of the other two, have you seen our handbook on care? No? Well, I only had one copy here. I wish you would just look at it and then you can write to us and get the information. We do have a handbook that might be helpful at this time, until we can get something better which we need.

MCKHANN: Let me ask you a little broader question. In your nursing curriculum...either of you can answer this...not limiting this question to Huntington's disease but in relationship to chronic neurological disease in general, how much exposure did you get?

LIGHTBODY: My exposure was very limited. I went to school-- I graduated about 17 years ago and my experience, my knowledge is extremely limited.

DUGAN: Well, I graduated 25 years ago and it was nonexistent more or less, in the curriculum.

LIGHTBODY: My only knowledge of Huntington's disease at that time was a young woman in the Kalamazoo State Hospital in Michigan. She was the only patient at that time who had Huntington's disease. And at that time, she was housed in what I considered a bad ward.

N. WEXLER: Would you say from your experience in caring for HD patients that there is a certain body of technique or knowledge for caring for neurological patients, especially with movement disorders, that would be useful to you as a practitioner to have learned? You had to do an awful lot by feel, I would guess.

LIGHTBODY: Yes. Skin care is a tremendous problem, both from the nourishment factor and from the positioning factor. If you try to restrain a patient in what you consider to be good positioning, often times he fights that and they develop skin abrasions and it just cycles. There's a lot to be learned about how to deal with this sort of thing.

N. WEXLER: A special curriculum could be developed then on this general area?

LIGHTBODY: As a student I was in the medical model and we need to get into the nursing model now and look at things just a little differently.

MCKHANN: Thank you very much.

The next three people that I have are Joan Masters, Phyllis Coyle and Rebecca Wilson.

TESTIMONY OF
JOAN MASTERS, R.N.
BOULDER, COLORADO

MASTERS: I did write a letter to the Commission, and one...my name is Joan Masters. I'm a registered nurse from Boulder. The only time I've ever become acquainted with Huntington's disease was with Dick Johnson, and I knew him for a year and a half until his death. And during that time, I think that the one thing that really impressed me the most was his suffering. He was a young man of 28 and he suffered many years before he came into the nursing home. It was the Good Samaritan Nursing Home.

The other thing I think that impressed me until his death was the fact that there wasn't enough knowledge known by lay people and by the medical people either. At that time, I was director of in-service and Mrs. Johnson--now, Mrs. Snyder--was very helpful in bringing enlightenment to us about Huntington's disease, which helped a great deal in nursing him. There were a few areas I think that could have been helpful in relieving his suffering and also in helping the nursing care; and that one area I mentioned in my letter, was the speech therapy.

I really felt that this was important because it got past the area, or past the time, when we could really communicate with Dick. And if he had had some help in that area before (it was too late then) we could have communicated with him longer. Just to give you an example, he wanted so badly to talk with us. He enjoyed people, loved people. And that was a big thing for him, and because he enjoyed this, he wanted to really tell us things that were in his mind. But the slurring of speech was so bad that he--if he had just said one or two words, we would have understood more than if he would say a whole sentence. And this would have been helpful.

Probably the biggest thing is the suffering. If there is some way of...within Dick's case too, he also developed epilepsy and I just learned from his mother that he did have some epilepsy at home. Now, whether this was part of the disease, I don't know. It usually is?--Okay. And the Dilantin did help him. Any other medications that were given to him just usually zonked him, like Valium or tranquilizers. (I probably am being repetitious.) But there just didn't seem to be anything else to relieve the spastic movements at all. Of course, along with the epilepsy--they were very bad.

I can't think of anything else right now, I think, other than what is in my letter. I'd be very glad to answer any questions.

Oh, there's one other thing--I had never heard of Huntington's disease before, and I also graduated from a very large hospital in Canada. It was a teaching hospital at McGill and I had never heard of it before. So this was a new experience for me.

MCKHANN: I'd like to ask you a question about that. We've heard on a number of occasions about the lack of training in this area. At what level...you've been involved in this kind more directly, I think, in training than other people who have spoken to us...at what level do you think this should take place? At the level of the undergraduate nursing training, or postgraduate, or both? And if people were to set up postgraduate courses and training relative to neurological diseases for nurses, would they come, do you think?

MASTERS: Yes, I think they would come, and I think it should start before postgraduate. I think it should be, because it seems to be more prevalent now and brought to the fore more now than before. I think that it should be in training as a nurse. And I think it's very important that more doctors and neurologists are brought to the attention of this disease because, as Mrs. Snyder can tell you, on more than one occasion, even a neurologist had no idea what to do, or in one occasion we know of, did not even come when he was asked because, we feel--and the doctor I spoke to before coming here, who was Dick's doctor, said he thought that it was probably because he felt he couldn't do anything and really didn't know, maybe, enough about it. So, I think in that area, it's not just the nurses, the doctors too.

TESTIMONY OF
PHYLLIS COYLE, R.N.
GREELEY, COLORADO

COYLE: I'm Phyllis Coyle. I'm an R.N. I'm also the mother of two children who are diagnosed as Huntington's disease. Their father was ill for about 15 years, was not diagnosed Huntington's, but was, first of all, diagnosed as a psychiatric illness and was placed, finally, because of finances and lack of space in other areas, in a state hospital. He was later diagnosed as multiple sclerosis and lived with that diagnosis until after his death; and with autopsy, and then the prevalence of other members of the family being also hospitalized, was then decided it was Huntington's.

This was not made known to me until our daughter was showing symptoms. She was pregnant with the second child at that time. That's when, actually, the diagnosis was made available to us. This illness had been a part of his family, their father's family. It had been hidden in the back room of the family home, and an old country doctor had taken care of them.

When their father was placed in the hospital, I asked then, "What do I do about the children?" And he said, "What about your children? Take them home and raise them. You got them out of the environment, and that's the most important thing." So that was kind of where we were at with counselling.

I was a nurses'aide at the time. We didn't have much money and we were never even given any counselling on disability, Social Security, or anything. So what we had we made ourselves.

Our daughter, like I said, was showing symptoms during her second pregnancy. After that pregnancy she was seen by a neurologist, who did diagnose her as Huntington's. At that time I asked him about some information on Huntington's and he gave me the Commission for

Control of Huntington's, the local chapter. And that was the information he had to give; nothing more than that. I called--no, I wrote to Mr. Wilson and from there we went. Our daughter was in three nursing homes--let's see, three nursing homes and one hospital --and it's usually ended up that they have called me to say "What shall we do with her? We don't know what the illness is. We don't know how to treat it, and we don't know what the prognosis is." And they've asked me for help, and I have offered the information that Mr. Wilson has given on in-service, and I'm sorry to say not one of them, to my knowledge, has taken him up on it. And they are all within the general area.

This is one area where I really think we lack, is in the education; not only of our nurses, but then within the institutions or within the facilities that we now have. Okay, to go on from there, our son was diagnosed in July. He's 25, our daughter is 24. He has one child; his wife and he are divorced. He is unable to work; he is now going to school, but his speech is very slurred; his gait is very poor; he falls frequently; he is still in his own apartment. We were unable to get any funds for him. He did have a small insurance that he was drawing on. Three years, and two attorneys, and he is finally going to get his Social Security Disability. He has went without food many times because he was too proud to ask, and we helped when we knew it was a need.

To our daughter's area of funding, the bills mounted and mounted and mounted until her husband was so overwhelmed with medical and hospital and doctor bills that he has sought a divorce because of it. He does not have any help with raising the children. He cannot even get, for some reason--and I don't even know why--money for child care while he works, and he does work steadily. Judy has been unable to get Medicaid through because of a bunch of red tape. Hopefully, once the divorce is over, maybe she'll be able to get that kind. Right now, Weld County is still funding her care, but that's very temporary. We buy her clothes--her stepfather and myself--and we provide her the extras that she needs. At the nursing home she's in now, she cannot even have a cup of coffee without paying for it.

This doesn't seem like a large amount to buy somebody's clothes or to provide the extras, but we also have two other children we're raising. They are still in school and they need a lot of things. So, I don't feel like I'm doing more than my share, but I do feel like we really have a desperate need for funding. The other thing we have a desperate need for, is for counselling; not only counselling as far as risk people, but for people like myself, because I need to go on working for my own benefit, as well as for my family's benefits, and if I don't have adequate help along the way, I can't function either. My husband, being my daughter's stepfather, also

needs some counselling along the way; if nothing else, to cope with me, and the other things that come up. Our little girls need help, not because they are risk, because they are not, but because they have a sister and a brother who have such a traumatic problem. I might add that my little children--my younger children--have probably been as much help to me as anybody has been.

Let me stop for just a minute. Our daughter and her husband did seek genetic counselling and did pay, I think, the \$40. And they then did decide not to have any further children, regardless of whether she would have been diagnosed or not, but they did already have the two. But there didn't seem to be anything after that, no followup; just, boom, here it is, do with it what you want to, and if you want a vasectomy, why, you can go to so and so and get it.

So, I want to stress again the fact that we need not only the genetic counselling, but the followup counselling. The other thing that I think we so badly need is treatment centers. We need them not only for Huntington's, but for the other related neurological illnesses, and I think particularly of patients with the Parkinson's or the other kinds of illnesses like that that could be similarly treated. They also need the counselling areas too. The funding for the children who are left without a parent somehow or another needs to be included in there.

There were several questions to the other nurses about whether or not they had any training--and some of them had graduated some time back. I just finished my training last June and as far as neurological illnesses, it was very, very sketchy. As far as Huntington's it was almost nil. And I'm not sure, except for the fact that I was rather verbal about my daughter being a Huntington's patient, because she was diagnosed during the time I was in school--I'm not sure that any of the students would have ever heard of Huntington's had it not been for that.

I had a friend who came here from the medical school at Miami last June and then was killed in the flood, but he was here during the time that we got Jimmy's diagnosis, and his knowledge--coming from a big medical school--was almost nil of Huntington's disease.

I think probably if I could suggest something that would just be, if nothing else, for literature available to the general public, as well as to schools. I'd like to see it from the junior high level, even before that, but certainly junior high and high school level. I think anything further than that is...I think you need to start down in the lower grades. I think then, of course, genetic counselling needs to be stressed more than it has been.

I think that's probably all that I have to offer today.

GUTHRIE: How do you feel about when you feel a child should be told that they may inherit a genetic disorder? Do you have any special feelings about that?

COYLE: I suppose, realistically, it should be quite young. But when you're the mother and you have something like that to do, it's very, very difficult. And I see as one of my prime responsibilities in life is to my three grandchildren. Somebody has to guide them now, and I'm not sure their father is going to be able to--he might. But if he does not, I think by the time they are 12 or 13, at the most, they should know that they definitely need to seek some help outside of dad or mom telling them what to do; that they need some professional help.

GUTHRIE: We hear, you see, some parents will say, "I don't want my child to live with the knowledge." Your answer implies that having the knowledge early in life maybe will help them strengthen their feelings about it, if they should have the disorder.

COYLE: Okay. I'd like to add a little bit then to that. Our son does not know he is diagnosed Huntington's. The doctors--both the neurosurgeon and the neurologist that consulted--decided that our son is very shakey emotionally right now and that he probably could not handle the diagnosis. I'm not at all sure that this is good. On the other hand I know, as a mother, I cannot tell him the diagnosis. If I tell the doctors to tell him and he does decide suicide is the answer, I'm not sure I can live with that either.

N. WEXLER: It doesn't seem that that ought to be your responsibility to face alone. And I think that's going to be a very strong recommendation from our Commission that there be help in that area.

TESTIMONY OF
REBECCA G. WILSON
DENVER, COLORADO

WILSON: My association with Huntington's disease began as a teenager, 12 or 13 years old, when my mother, Mrs. Margaret M. Wilson, started showing symptoms of H.D. The symptoms that I noticed were changes in mother's behavior. She became more and more moody, suddenly angry--then regretting the anger. Mother at this time began spending more and more time at a nearby shopping center and being away from home as much as possible. This was a distinct difference from the good-natured and concerned homemaker that mother had always been. I now think that the times she would spend away from home were her attempts to prove to herself that she was still a person competent enough to be with other people. It was also at this time when Mom began losing her sense of balance. She would stagger and was unable to walk in a straight line. People assumed she had been drinking. In our family, we began to lose friends because of this.

During the next two years, I do not recall many distinct changes in my mother's condition. She continued having the same symptoms, but perhaps the progress of this terrible disease was slowed.

In 1967, Mother's affliction due to Huntington's disease became worse. All the symptoms were exaggerated and intense. She would be very depressed and very angry and would scream and fly into rages. I remember there seemed so much conflict within her then, how hurt she was that she could not control herself or her hurting us. These were some of the harder times, and how sad, yet almost easy to emphasize with how she must have felt.

We sought help and information from Fitzsimmons General Hospital. One doctor, Dr. Madison, explained some matters about H.D. that I had not already known. At this time I was aware of my at-risk position. In general, Dr. Madison was unable to offer help as to how to cope, what the progress of Huntington's disease would be like, or what the hopes for a cure were. Dr. Madison was one of the very few doctors at Fitzsimmons who had knowledge of H.D.

In autumn of 1969 it became necessary for me to quit high school to help care for Mom. The staggering had increased and Mother was often violent. It was at this point when Mom would pull a kitchen butcher knife on me. After a few minutes she would give me the knife, cry, and tell me she loved me. I am not belittling my mother here. Incidents like these are examples of the devastating effects H.D. can have.

It was necessary to have someone else to help care for Mom; we hired a nurse for the time dad worked. This was expensive and the nurses did not stay with us very long. Unable to cope with the situation, I had been sleeping approximately 16 to 20 hours a day, and did seek psychological help at Fitzsimmons General Hospital. The psychiatrist prescribed tranquilizers which I continued for a few months and do not use now. My mother was admitted to Fort Logan Mental Health Center in Denver for a short period. As family, we agreed she was still mentally competent.

It is difficult to say when mother began losing part of her ability to speak, though she would repeat herself 40, 50, 60 times and forget what point she was making. Today, she doesn't or cannot speak many words at one time, however, she knows what she wants to say, and what she needs. Some people can understand what she is saying; my father and I seem to understand best. This communication reminds me of a child learning to talk, whose parents understand better than others what the child is trying to express.

The last seven years have seen Mrs. Margaret M. Wilson (Mickey, to her friends) grow steadily weaker. Father has told her that when

she cannot control her bowels that she will have to go into a nursing home. My father is a man with strength, patience, and love. My mother also has strength in her spirit and attitude. She still loves, cares for people and life; is interested and aware; and she is happy to be at home--not bitter because of H.D.

Our daily life seems fairly ordinary, though it is somewhat unusual. When mom sits she cannot sit for very long; when she walks she appears to be about to fall over, and often she needs slight assistance. The atmosphere at home can be nervous or tense. Either my father or me is with mom at all times; in the past it has been difficult for others to stay with her. Our schedules are in part determined by mother's sleeping habits. There are many times when she doesn't sleep for three days or more. My parents go out to a few places where people know Mom: bingo once a week--the Moose Club built a carpeted area with a deep cushioned chair--and Mom and Dad go for ice cream twice a day or so. Mother can feed herself with a spoon, but she spills about half of what she eats. A friend made bibs for her. Mom still smokes cigarettes occasionally; this seems our weakness. Between spilling on her clothes and occasional burns, Mom goes through clothes rapidly.

Two notes: 1) Mother is almost always moving, and restless, except when she is sleeping, and then she does not move unnaturally.

2) We try very hard to communicate with Mom and encourage her to communicate with us. I believe this to be of utmost importance. Perhaps, I believe this helps all of us to be the best we can.

In a sense my association with Huntington's disease began when I was born. It may not end when I die, but live on in my children or grandchildren, if I should have children. At this time, I feel I will not--the chances of helping eliminate H.D., itself, by not having children, and the possible guilt in conceiving with Huntington's disease affects this reasoning. There is a more positive side also. Forty or so years of life can be worth the H.D. risk, and there must be hope and research to eliminate Huntington's disease. There are many people willing to work toward this goal: concerned doctors and medical people, people associated with H.D., the general public and me.

In writing of children, I didn't mention a husband or father. The strain of Huntington's disease upon any relationship, especially a marriage, can be unbearable and heartbreaking. If I am to develop H.D., this would be a difficult burden to ask of someone I love. And what if I marry, have children, am developing Huntington's; do I say to my husband, "Please take care of me and, darling, let the children

know I love them, and try to prepare them for the possibility that they may develop H.D.?" Perhaps, or I could marry without telling my husband of the risk of Huntington's disease, This I would prefer not to do.

Socially, or as a part of my personality, I often feel somewhat withdrawn. I know I get depressed and quiet at times, and I think this is partly due to my association with H.D. I may be slightly bitter, or feel sorry for myself. I recognize the very real possibility of developing Huntington's disease, and see it reinforced every day. I can, and with work will be more positive and hopeful, but I do not wish to deny or lie to myself.

For anyone who develops Huntington's disease, if no cure is found, no controlling agents found, no better methods developed--what choices with a life of being a, often-called, vegetable? Years full of frustration, depression, decreased social contact, loss of pride. Nursing home costs increase, insurance is nonexistent or unreasonable, at-home care is often impossible, and frequently the "breadwinner" is the one afflicted with H.D. There are four choices: nursing homes, at-home care, state institutions (often overcrowded and understaffed), and suicide. Suicide seems the most defeating and perhaps amoral, yet many people have already seen it as their only alternative.

My future, as well as the future of thousands, millions of people, lies, at least in part, in this hearing. The beginning of new hope and ways for a better life? We hope and pray so.

The following are problems or areas I think worthy of mention. Problem: Care of the diagnosed H.D. patient. Often the patient with Huntington's disease needs specialized care, both physically and personally, psychologically; as a fellow human being. The course of the disease itself can or does progress at different rates and manifests itself in different forms for each individual affected. H.D. patients can be very active--this is often a problem with nursing care and institutions. Very often, the H.D. patient is mentally competent, alive, and aware as a person. Perhaps, therapy could help the victim of Huntington's learn to cope and express--be more productive, less hopeless.

Recommendations: First, I would like to recommend that, if possible, Elisabeth Kübler-Ross be suggested as a qualified person to contact regarding the personal, psychological care of H.D. patients. Through my college psychology classes I have been impressed by her work with terminally ill persons. Secondly, training in the care of H.D. patients could be offered to nursing staff and institutions. And thirdly, at-home care might be helped by organizations such as CCHD, clinics, and counselling.

Problem: Lack of a professional informative source. Without some professional knowledge as what to expect and how to deal with various aspects of Huntington's disease, there is very little that can be done to help the affected member and family physically, psychologically, and financially.

Recommendation: (1) The idea of centrally located clinics with the services offered in close proximity is a concept recently presented to me. For the "drawing board," it seems practical and logical. (2) Other services that might be offered include family counselling, genetic counselling (I went through a genetic counselling program), and some type of counselling with social, psychological or psychiatric services offered. (3) There should be a listing of doctors available, nursing homes and alternative care. Also, if possible, there should be someone with information concerning the various financial aspects of H.D. (4) Perhaps there could be suggestions as to when, or how, to best tell threat-risk children of their possibility of developing H.D.

Problem: Finances. Some of the financial problems associated with Huntington's disease that I see are as follows: (1) Often the "breadwinner" develops H.D.--the family may be unable to support itself. (2) There can be many medical-related costs which are increasing--doctors' fees, treatment costs, nursing home costs, expenses for live-in nursing care, at-home care bills, et cetera. (3) Little or no insurance coverage.

Recommendations: (1) Perhaps Federal or state aid or allowances for H.D.-related expenses. (2) Reasonable or low-cost medical and counselling services.

Problem: Huntington's disease has not been eliminated.

Recommendation: Research should be continued, encouraged, and communicated. Research is very important and encouraging.

Thank you for the opportunity to express my thoughts and opinions concerning Huntington's disease.

GUTHRIE: I love all of your recommendations. I think that they are all urgent and I think that I am glad that somebody has stressed the need for patients to function as long as possible to the best of their capacity and not be judged on what they might look like five years from now, but what they are today.

COYLE: May I add something to that? Our son is in school and because of another problem that he had, he's been going to school under a rehab program. They have recently found out through his psychologist that he is diagnosed Huntington's. So, the decision has

been made, however, I'm not sure yet if it's going to be carried through--where I'd like to see it not carried through--the decision has been talked about now of rescinding his rehab funding for finishing his school. He has one full quarter after this one to go for his Bachelor's degree, and I think that's terribly unfortunate because he needs these goals to work toward, and I'm not sure if he will be able to use them. But I don't know that he will not be able to use them.

GUTHRIE: And he needs the opportunity to prove whether he can or not.

WILSON: May I say something? When you were talking about how much nursing is known, I just completed my first year of college, and we have a nursing program and I have quite a few friends in nursing. I mention Huntington's disease to them, and I'm telling three out of four of them what it's about and I know two women who are graduate--they're going back there for additional training--and the most they know about Huntington's disease is that it has chorea movements. They have no idea of the mentally competent, aware state. I mean, it is just, you know--or how--or no recommendations for better care.

N. WEXLER: If they cut off the funding, let us know.

COYLE: Okay.

N. WEXLER: Seriously.

MCKHANN: The next three people on my list are D. A. Link, Dr. Carter and Dr. Maul.

TESTIMONY OF
JACK L. CARTER, PH.D.,
PROFESSOR & CHAIRMAN
DEPARTMENT OF BIOLOGY
COLORADO COLLEGE

CARTER: First, I would like to thank the Commission for allowing me to race up here in a rainstorm from Colorado Springs to place my two cents in this very important discussion. I'd like to speak from the point of view of a person involved in education and reaching out to society at large. And I'm going to repeat a few statements I've placed in my letter and maybe add a couple points and then step back and allow people to ask questions or maybe discuss it further.

I'm also suffering a spring allergy--you'll have to suffer through that a little bit.

It is my opinion that if we were to survey the people passing almost any street corner in America, nine out of ten people could not define or describe the causes or effects of Huntington's disease --or for that matter, most of the 10 most common genetic diseases. For this reason, I would like to direct my statement to the item number 4 of the list of major objectives for these hearings, and to point to the need to generate interest in Huntington's disease and other genetic diseases through a national educational program.

Huntington's disease, alone, causes a great deal of suffering for a relatively small group of people, but if we combine those persons affected in one way or another with Huntington's disease, with those persons affected with other well-understood genetic diseases, we may then have a large group of people who could have a powerful voice for educating and communicating with the total population.

Down's syndrome, sickle cell anemia, Tay-Sachs, Turner's syndrome, Huntington's disease, and a number of other human genetic diseases in the total affect a large number of people, resulting in lost lives, countless years in hospitals and health care facilities, and requiring the expenditure of millions of dollars in private and public funds. This total problem is worthy of the attention of the Congress of the United States, the medical profession, the health care specialists, but most importantly, educators at all levels. Nothing distresses voters, by the way, more than to read where Congress has voted a large sum of money to fight a disease or diseases of which they have never known a single person to suffer.

This, then, is the problem. We must reach out through a variety of media to all levels of our society and improve their knowledge of these diseases--inform them of the role of medical research in fighting these diseases; help them to understand the need for genetic counselling centers in communities and schools; and relate the closeness of these diseases to all mankind.

If we, as a starting point, devote our time and limited funds to education and, consequently, stimulate interest in research and counselling, it will then be possible for Congressional leaders to attack these problems on a national scale. And at this point they would not have to fear reprisals for their supporting of the research and counselling centers.

I would suggest the educational program work to reach students in our schools and colleges, but not be allowed to center on this group of people. Much of the most important education that takes place in the United States today occurs outside the traditional educational institutions--television, radio, newspapers, magazines all play a major role in education, and especially adult education. It is very important to keep in mind that this adult population also pays the bill for most of the medical research conducted in this country--this must be an informed audience.

Now, let me describe a model that I included in the letter just as an example of how I think we might go about reaching the larger segment of our society. If the Commission were to develop an excellent 16 millimeter 60-minute color film to be presented on public television, describing Huntington's disease, and if teachers and parents were notified well in advance when it would be shown, millions of people would be informed of Huntington's disease perhaps in a single evening. Now, to carry this plan a step further, if the schools and colleges were notified well in advance of this showing and told the film is public domain, and were encouraged to copy this film from television onto their own tapes for continued reuse, people of all ages would see this film several times over the next several years. This program could, if well presented, literally place pressure on Congress to vote funds to fight this and other dreaded human genetic diseases. I give this only as an example.

I brought along...each week we receive from...on Channel 8, which is the educational television station in Colorado Springs...we receive a printout on campus of all the programs that will be shown. I go over this with our department, discuss it with faculty, and we have certain programs copied. And we use the Nova series over and over in general biology and beginning zoology classes. We use many programs in this way--The Ascent of Man, the Bronowski series--and I use these as examples of how I think we might go about this. There are many other methods in a variety of materials that could be developed and used in a national educational program. I would urge the Commission to identify these, study them carefully, and develop those that can provide the greatest dividends in the shortest period of time.

GUTHRIE: From what you're saying, though, I'm assuming that you would also approve the idea of perhaps a program that would show how these diseases are related to each other, not just specifically about the one disease?

CARTER: Certainly--no, that's the idea here. I used that model, but if we would, say, take 12 programs and start out with the basic genetics that are necessary for people to understand all of these diseases, then follow through with examples from counselling centers, medical schools, where people are treating and working on research; and some of the kinds of things I've heard here just in this short period this afternoon would be appropriate for that showing, I think.

N. WEXLER: I think that there's precedence--actually, the show on H.D. in England which was presented and got excellent response and someone reported in the Seattle testimony that everywhere she went people knew about this disease. I think that--a statement and a question--one of the things that we want to communicate, which, I have been told, is very difficult to get across on public TV--is how do you get across basic concepts, genetic inheritance, and, even more difficult, how do you get across basic concepts of basic biological research and

research particularly in brain and brain functions? HD involves really both realms. How do you increase support for basic neurological research? I've been told that it's difficult to get a jazzy TV show on the brain, Question; How much would such a thing cost?

CARTER: Well, I've talked to others about--not just this, but other television programs, designing and putting them together. Something like the Bronowski series ran over a million dollars, about a million and a half a showing, and it's very expensive. But I think that it would pay dividends in the long run.

Now, back to the first question about the teaching of basic concepts, and a very important part of teaching genetics is teaching the ideas of probability. I think anyone who has played around with those ideas knows how important they are to our society today, not only in genetics, but environmental and energy problems in a lot of ways. I think that you have to go over some of these things we know several times so that you do have to quite often teach, reteach and present them from a little different angle. But I think you could--if you had one program that dealt with these concepts--and then in the beginning just review them, hit as they do with number theory, and things like that in the Bronowski series. You can come back to those again and again, and before long--aha--that's the same as it was--Down's, yes; sickle cell, yes; Turner's, yes--okay, I get the picture. And I think that might be the attack or the approach you may want to follow. I'm not any authority.

MCKHANN: Dr. Carter, one of the points that was brought out to us earlier this morning, and we've heard from other people as well, is the lack of teaching in human genetics. Do you have any thoughts on how that might be improving?

CARTER: Well, we recently conducted a little survey of a mid-year conference up at Greeley, and we've tried to pull some data on this. I think if you ask most high school and college biology teachers--college teachers teaching the beginning courses--they will say they are covering human genetics. But quite often they're teaching the principles in that case, I believe, without talking about the various diseases. In other words, they do teach the human genetics--basic genetics--but they never deal with the individual diseases and the variations in these diseases in those classes.

Now, perhaps--and I think they would probably say--that we can't cover all of these. You see, well, there's no way that we could cover all of these and we try and teach genetics first; then, after we get through drosophila and everything else, we get to human genetics and I think it gets cheated--I think there's something to that; it does get cheated.

GUTHRIE: I would just like to ask you--we musn't forget that Huntington's is also a brain disorder and I'm just wondering, do you know anything about anybody doing any education or teaching of how man's brain functions as part of, also, an educating curriculum in the lower grades, or possibly high school?

CARTER: No. I learned through another teacher in a college in Iowa--was the first time, not too many years ago, that there is a brain involvement in Huntington's; so that it's not taught, generally, in our schools.

GUTHRIE: I thought that might be another opportunity for education.

CARTER: Right, it is.

GUTHRIE: Going to man's brain might be more exciting than going to the moon.

CARTER: Right. We should try to reach that point, I think, in our thinking.

MCKHANN: Thank you.

TESTIMONY OF
KESTER V. MAUL, M.D.
CHIEF MEDICAL CONSULTANT
REGIONAL MEDICAL OFFICER
BUREAU OF DISABILITY INSURANCE
SOCIAL SECURITY ADMINISTRATION

MAUL: I'm Dr. K. V. Maul, Regional Medical Officer for a Bureau of Social Security Disability Program. I have today with me a young lady, Jackie [McDow], who is a field representative for Social Security and thought she might have some questions come up that I'm not...that are out of my area, she might be able to answer them. She also has with her several pamphlets that I'm sure she'd be glad to hand out that might give you a little bit of information of how Social Security might play a part in Huntington's disease.

First off, I should say that the Social Security Disability Program is a unique program as far as disability is concerned. It's unlike most other programs--for example, it's an all-or-none program. Now, most of you are familiar with the VA, State Comp --that has a rating and a percentage-type thing--private insurances. And Social Security, after the eligibility criteria are met, it's an all-or-none. You're either allowed benefits or denied benefits. There is no provision in the Disability Program to show any particular favoritism--maybe that's not a good word but there's no particular disease, or series of diseases, categories of disease are singled out for any particular interest. They're all looked at under the same very basic disability criteria.

In the short time that we have to discuss this, I can only hit on a very few of the high points because it's a very complex subject. But I would like to emphasize a diagnosis, in itself, is not sufficient to fulfill the criteria under this program. It has to be supported by certain objective findings. Actually, Huntington's disease makes up a small percentage of the cases that we evaluate under the Social Security Program. The conditions that we look at are broken down into 13 major categories.

For example, we have the musculoskeletal system, cardiovascular system, pulmonary system, gastrointestinal system, mental diseases, malignancies, and neurological section. Under the neurological section, we have many conditions under there. For example, we have strokes, we have multiple sclerosis, we have muscular dystrophy, we have polio, we have Friedreich's ataxia, and, of course, Huntington's.

All of these conditions are looked at by an evaluation team in a state agency. This team consists of physicians, medical specialties in various areas and what we call adjudicators or examiners. They're technically trained people that accumulate and look at some of the data, and interpret some of it that is of a non-medical nature, and finally, if sufficient information seems to be available, a decision is made. Now, there are a number of appeal steps, because if a person is denied benefits and thinks he should be entitled to it, of course, he's not very happy about it. And there are several steps that can be...he can follow to appeal this process.

To condense this thing in this short a time is a pretty difficult job, but I want to leave a few important points, if I can. I mentioned that a diagnosis by itself is really not sufficient. Many people, I hear constantly saying, well, I know so and so that got benefits and he's drawing 100 percent VA Disability, or state comp, or whatever, but he was denied Social Security Disability. That is true--it can be true. I've learned from years of experience to hesitate to stick my neck out and make an explanation if I don't have all the facts and have the case in front of me. Because what may appear on the surface to be two identical cases, when you get into it, you'll find that there's some ramification that makes a difference.

Anyone who believes that they may be eligible for Disability under any condition is certainly entitled to apply at any district Social Security office, and be interviewed and submit an application; that information is finally funnelled over to a state agency. There are many district offices in each state--branch offices. There are several in the Denver Metropolitan Area. There is one state agency in each state where the information is finally funnelled into and the decision is made.

I mentioned that Huntington's, which, of course, interests this group here, I'm sure--and rightly so--actually makes up a small percentage of the cases that we see. They're important to the people

that are involved, I'm sure. The physician who may be the treating physician, or any treating source that the person may have gone to-- I want to make it very clear that he does not make the Disability decision. He furnishes information which is then funnelled into the state agency that I mentioned, because a condition may be severe enough in and of itself to meet the criteria. Another individual with the same impairment, because of the vocational factors that sometimes come into play, can weight the decision one way or another.

It is, as I mentioned, a rather complex type of evaluation and a lot of information is necessary and a lot of time and effort is spent with this team that I mentioned, in trying to come up with a fair and equitable decision.

There has recently been quite a push by one of the subcommittees of Congress to try to insure uniformity over the country so that one area, one state, or one particular area does not go overboard one way or the other. And there has been a General Accounting Office audit recently with much emphasis put on training all over the country and all Social Security agencies for more uniformity in their decision making.

Now, by the very nature of the process in trying to evaluate disability, it's not like mathematics or physics, where it's a precise science--believe me, it's a tough problem. And it will never be as accurate as those sciences I mentioned, but everybody is trying to do the best they can and I think reasonably--that they're doing a reasonably good job.

I think that--unless there are some questions, I think I'd better stop right there.

GUTHRIE: I would just like to ask you, knowing the prognosis of a patient with Huntington's, I'm assuming that they would have the opportunity for a continuing evaluation if they were turned down the first, the second, or the third time?

MAUL: That's correct. The first step in the appeals process--when a decision is made and the applicant is notified of the decision, which in that case would be he would be denied benefits, he has the privilege of asking for reconsideration.

Now, to enhance his likelihood of receiving benefits, if he has some information that is newer than what was originally submitted, or some different information, or anything that he thinks may throw a different light on it, of course, that is helpful. Also in the reconsideration process, a completely new team of evaluators looks at the information so that it gets a different look and is not biased by what the previous team has indicated. So that the person always has a privilege of reconsideration and, hopefully, if he has additional information, it will certainly be considered.

There are two main aspects as far as the medical situation is concerned in evaluating any disability. First, is the level of severity. Now I mention again, the diagnosis--sometimes the diagnosis itself--the name of the condition has an ominous sound--cancer, for example, always scares the pants off everybody, and it should. But there are some cancers that are much more serious and fast moving than others. There's a duration factor. In other words, a condition either has...it has to have reached the level of severity--a certain level of severity. It also either has had to have existed for 12 months or longer--it's not a short-term type of deal. We have a lot of serious problems that would prevent the individual from working for a period of time, but this is a longer type of thing.

Does that answer your question?

N. WEXLER: If a woman has been working in the home as a housewife and a mother, how does she get Disability benefits?

MAUL: Well, in the first place, there are two big divisions. There's a Title II and a Title XVI, and we could take all day on that and I don't want to get too far off on that, but when the old original Social Security Program started back in the middle '30's, it was designed strictly for people to receive money after they reached the age of 65. They paid in and their employer paid in a certain percent. That went along for a number of years, and then in about '54, I believe it was, the first disability provision came into play. This was a so-called freeze. I won't explain that, but it essentially enabled a person's average level of earnings, upon which his ultimate income would be based, or his benefits would be based, he wasn't penalized for no earning or low earnings years. But then they came on and then added a number of provisions and the disabled widow came into play, and the children came into play, and the disabled widowers. And in Title XVI, it's not based on persons having worked in covered employment; it's a needs program or an income and resource program. It came in in January of 1974, I believe, so that it has just grown by leaps and bounds; and then even the Black Lung Program came in to muddy the waters further. So, it is possible for a woman, a housewife, who has never worked--she might be covered through her husband who would have worked under covered employment.

N. WEXLER: We also heard just Friday in Los Angeles about a woman with HD who testified very eloquently that, of course, her work experience had been somewhat hampered by her disease. When she finally was no longer able to work, she was missing one quarter only, so that she was not entitled to any benefits whatsoever, although, I think, between her and her husband, they had contributed at least \$50,000 to Social Security but they were eligible for no benefits whatsoever--for one quarter.

MAUL: You know, it's like a lot of things--there's a lot of people that's slipped through the cracks in this thing and sometimes--

N. WEXLER: That's a big crack.

MAUL: Sometimes people get allowed that we know darn well shouldn't, but there's that little quirk in there that makes them eligible; and there's other people that you have to deny--that you feel bad about it, but that's the way it is.

N. WEXLER: Well, maybe that doesn't have to be the way it has to be, and that's why this Commission is here. We want to know how to recommend--what recommendations to make to alter those eligibility requirements.

GUTHRIE: In fact, I'm going to be pointed and ask you, because we heard this woman's testimony and it was so sad having paid in \$50,000, unable--have you any tricks that you might tell us? How can we help this lady get something out of what they have put in?

MAUL: The only way that I know to answer that question is that as long as the law says you have to have so many quarters--it like if you put 95 cents in the bank you only get credit for 95 cents. The only way I'd know is that if they feel that that's unfair then they...the law would have to be changed. But you wonder, well, where are you going to draw the line? They decided that a person should have so many quarters--20 quarters out of the last 40 to be eligible under Title II--and unless you change it, that's the way it is.

GUTHRIE: Maybe we can change the ratio, huh? After all \$50,000 and missing one quarter, well, we can change that.

MAUL: Sounds too bad, but that's the way the law says it is.

GUTHRIE: We have a lot of work to do.

CARTER: Perhaps if the law was written so that the husband and wife together--the total quarters--I mean, I've...it doesn't seem like it would be impossible to work that out--I don't know.

N. WEXLER: I think that also discriminates against single people. I think we need a new definition.

TESTIMONY OF
D. A. LINK, ATTORNEY
GLENWOOD SPRINGS, COLORADO

LINK: My name is Link; I'm an attorney and I practice law on the western slope of this state. I have no particular medical qualifications to testify here. Although my profession gives me quite a

bit of opportunity to speak before groups of people, this is a bit unnerving for me because I'm not accustomed to talking about this with anyone, much less, at a public hearing.

My mother has Huntington's disease and I am at risk. And my father and sister testified before this Commission in Los Angeles ...lawyers aren't supposed to be this way.

GUTHRIE: Let me help you out a little bit--let me just let you calm yourself and tell you that your father and your sister were most eloquent in LA and your sister said something that is vital that you might like to hear. Your sister came in late and did not hear your father's testimony. So having come in late she said, "I don't know what my father said, but I would like to say what I think." And she said that if she had Huntington's, she didn't want to live.

And as luck would have it, sitting right next to her was a man that I knew personally, who at the age of 65 had had Huntington's for many years, and was a violinist, and he was about to testify. So, I said, "I'm awfully glad you got here and I want you to listen to the man sitting right next to you, because he's going to tell you some things about Huntington's that you don't know." And when he finished, your sister turned around and said, "I have never seen any other patient and it is an amazing experience for me to see a positive approach to this disease."

Now, with that, perhaps, you can speak a little more clearly.

LINK: Well, my mother is in the advanced stages of the disease at this time. I would like to point out that I do not have any particular expertise with the legal problems of Huntington's disease patients. I may have firsthand experience in the future, but as a lawyer, it's not very difficult for me to imagine the problems that an HD patient would have with both the civil justice system and the criminal justice system, particularly if they have not been diagnosed HD, and in view of the fact that often times the symptoms have their onset well ahead of time--before they're diagnosed.

I would like to, before I forget--I've never met you, Mrs. Guthrie, and I assume that's who you are from the nameplate--but I'd like to thank you for all the work you've done with the Committee to Combat Huntington's Disease. I wish there was some way you could be repaid, however, if satisfaction for a job well done is any kind of measure, I hope you're a very wealthy woman.

GUTHRIE: I'll tell you right now, I am, and I want you to know we all have to work together. And that I don't mind being the starter but, boy, I want a lot of helpers, right now.

LINK: My recommendations cover three basic areas, the first of which is research. From what I can tell, this area needs one heck of a lot of research--and research, in my opinion, in three particular areas. First of all, research with treatment. I am not fully aware of all the progress that's been made with respect to treatment, and I'm sure I won't live to see the full extent of the treatment possibilities, but others that follow me, I hope will. I hope that certain...

GUTHRIE: I'm going to stop you right there--forgive me. You are going to live to see treatment. I won't say cure, but I believe as a young man, you're going to see treatment. We're on the threshold. I can't let you just not believe it.

LINK: Well, I think, that's one particular area that should be focused upon first as far as the research is concerned.

The second of all, and perhaps the most important to me as an at-risk, is the detection of the disease. As far as I can tell, very little progress has been made in this regard and I, personally, would like to know. I'm sure that the initial discomfort or pain of knowing that I will be diagnosed as one or the other would be very great, but if friends and others are available, I'm sure I could be weathered through. The fear and uncertainty, to me, is not worth the knowledge. I think, most of all, professionals could stand to benefit from tests, which could definitely tell one way or another, regardless of the age, whether or not an individual had HD.

The individual who diagnosed my mother knew very little about the disease and just kind of unloaded it. And for years, my mother had ...looking back now I can tell...had manifestations of the disease. And my father, being a very conservative and well-disciplined man, oftentimes joked with her or ridiculed her about her symptoms. And I, personally, would have liked to have seen him spared discomfort when he discovered what he had been ridiculing.

Second of all, I think tests which would allow early detection of the disease would enable those of us at risk to better understand and cope with the reluctance of others to share the burden and the uncertainty. I had a vasectomy at age 21. This is a decision I'm quite sure was right, but one I'm still having difficulty living with.

I was married for two years before my wife decided that she could no longer live with the uncertainty. And up until three weeks ago, I had a fiancée who went home to visit her folks for a week, and also decided that she'd better not take the chance. In spite of the way I'm feeling, I really can't blame them, nor can I blame her parents, who were very instrumental in influencing her decision, because it's difficult to realistically expect someone to share the kind of uncertainty you feel, wondering whether when you drop a glass or have a

sudden bit of anger, whether it's the onset. I think other individuals can take unfair advantage of the uncertainty as well, because I had this vividly demonstrated to me shortly before I got married. I became aware that my mother had been diagnosed and I was considering getting life insurance for myself and protection of what I thought was going to be my future family. Two insurance companies turned me down when they learned that my mother was diagnosed HD. The third, I made the mistake of telling him that the previous two had turned me down and he persuaded me that I might never be able to get insurance unless I accepted his policy, which I did. Looking back on it, it was a stupid thing to do, and I was fortunate to be able to get out of it, but I think it's a fair example of the kind of over-reaching that can take place.

The third area that I think the research needs to be done in is in the area of cure for this thing. Perhaps that will come with the research in the areas of treatment and detection, but ultimately, the objective has to be the cure. What little I know about genetic diseases, I doubt, I don't have much hope that there will be a cure, but I would hope that appropriate attention would be focused in this area.

A second recommendation I would have relates to education, particularly the education of professionals. As I said before, the importance of early diagnosis to my mind cannot be overemphasized, especially in relation to the associated emotional and financial costs.

The first gentleman in our group, I think, pointed out very well the fact that the public needs to be educated in this regard. I would hope a very strong public education program could take place, if for no other reason than to perhaps get some public support financially for the efforts to focus on HD. Education for the families who are immediately touched by this disease seems to me to be sorely lacking. I think education on how to cope with this disease, how to care for a member of the family who has been diagnosed HD is particularly important. The victims themselves, I should think, should not be overlooked with regard to education.

And my third recommendation would, of course, be financial aid in whatever form possible. Financial aid for research and education, facilities and treatment personnel. My family was--is--has been particularly fortunate with HD because my father has done fairly well in his business and we are not immediately threatened with tremendous overwhelming medical costs; that situation could change. I know a lot of families are not in the position that mine are and that seems to be most unfortunate and unfair. So, I hope that, if nothing else, this Commission can put forward some recommendations to Congress for effective financial aid.

That's all I have to say.

GUTHRIE: I have to comment--when my daughter was 17 and "Alice's Restaurant" came out, and the public knew that the Guthrie family had Huntington's, we had a little meeting. And Nora turned around to her brother and put her arm around him and she said, "Jody, wouldn't it be terrible if you lived to be 40 or 60 and you waited for Huntington's to come and it never came and you never lived?" That's a little bit of thinking for you.

MAUL: We'll leave those pamphlets, I guess, over on those tables for anybody that Jackie has that might want to pick up one.

MCKHANN: Is Dr. William Powers here? If not, then the next people that I have on my list are Phyllis Clements and Beverly Beauvais, and John Arcotta. Are they here?

TESTIMONY OF
PHYLLIS CLEMENTS
PUEBLO, COLORADO

PHYLLIS J. CLEMENTS
PUEBLO, COLORADO

MARCH 21, 1977

"A FATE WORSE THAN DEATH"

This is the story of how Huntington's disease has affected the lives of Tom and Phyllis Clements, both age 45, and our four children, Lynda, 24; Dan, 23; Bill, 22; and Mike, 20. Through contact with other Huntington's families, we know we are not alone, and yet we seem to be an island, as each victim and family have such individualized reactions and needs.

I will try to relate some of the problems and heartaches our family has personally experienced.

At the time of Tom's diagnosis, he was just finishing 20 years in the Marine Corps. I had a good income job, we had four lovely children in their early teens, a beautiful, big home and lots of plans for the future. We were completely unaware and unprepared for Huntington's. Tom is adopted.

Tom began having trouble with his coordination and developed a stagger-like walk with a slight swaying motion when he was standing still, of which he was completely unaware. His superiors began accusing him of being drunk on duty. At the time, he was also a Little League coach, and the parents began making the same accusation. His superiors believed him, but his co-workers felt he was faking some strange illness in order to get out of his share of duties or to receive better benefits upon discharge. After several accusations and just as many denials from Tom, he was sent to Camp Pendleton Hospital for diagnosis. That was in November of 1968, when Tom was 37.

The doctors at first were at a loss, but finally a neurologist saw what he thought were signs of Huntington's disease. His adoptive parents knew who Tom's biological family was; in fact, lived on the same block and had adopted Tom when the family of three children split up. His natural mother had been hospitalized at Colorado State Hospital in Pueblo, where she had died of "some kind of nervous disorder." The doctors sent for death records and they stated that she had died of Huntington's chorea, confirming the California doctor's diagnosis.

When we were told Tom had Huntington's, at first it meant nothing to us, but it wasn't long before we realized this was

not just any kind of ordinary disease. Literature was hard to find, and the doctors did not have much more to say other than that within four years he could be a vegetable, the disease was hereditary and statistically speaking, two of our four children would also be victims someday.

I remember being totally bewildered. Tom was stunned. The children took the news with very mixed emotions. Just entering what should be one of the best times of their lives, they had to face the fact that they too could be victims. We took them to genetic counseling and kept them involved, but as they saw their Dad degenerating in front of their eyes, they became at first confused and then, as time passed, more and more embittered. Their plans for the future have become marred by the fact that there may be no future for them to look forward to, except to end up the way their Dad has.

To this day, they live with this shadow over them. But they are trying to accept what life has to offer. They seem to have faith that something will be done before their children will be affected. Two of our children have decided to gamble on the future. Our family now includes two beautiful and bright grandchildren, who are also at risk: Janet, 5, and Brian, six months. Their innocence and spirit have been not only a joy to all of us, in spite of our fears for them, but a symbol of hope for the future.

Fortunately, at the same time of Tom's diagnosis, Margie Guthrie was founding the California Chapter of Huntington's disease. We were among the first to join. There we not only received much needed information and encouragement, but a small feeling of relief that there was other people we could relate to, who also had a tragic future to look forward to. We were in contact with people who were learning to cope with the disease and had hope for the future. We were also in contact with doctors interested in and dedicated to researching the disease. We were available, whenever possible, to give them blood, skin, or whatever was needed.

Tom volunteered for a brain biopsy, not knowing what the after effects of the biopsy might be on him, but hoping they would find something to help future victims. He knew whatever they found would be of no help to him personally. We received black and white prints and a full report on the findings, including the report that the researchers had been able to keep enough brain cells alive to share with other research centers.

At first the children were eager to help, too. Then they were asked to give a small sample of spinal fluid. We were told that there would be no aftereffects, so we gave our written consent. The doctors took approximately three test tubes full from each one, plus some blood samples, and sent them on home alone, 60 miles away. The children suffered severe headaches, and we had four very sick children who weren't back to normal for a month. There wasn't any follow-up concern with how they had reacted to the testing. They also had been told that they would be kept advised of the findings and progress or lack of it from each test they volunteered for. We heard nothing. The children felt used. Like laboratory guinea pigs whose only function was to provide some researcher with more data. It embittered them so, they have refused to get involved in any more research projects.

After Tom's problem was diagnosed, he was discharged from the Marine Corps. He was advised not to drive and his license was subsequently not renewed. Though not highly impaired at this time, he was unable to find other employment. No one wanted to hire him when they saw the reason for his discharge. It took over 18 months to get his Social Security and Veterans benefits straightened out. They were unfamiliar with Huntington's and couldn't understand why he couldn't work. They required positive proof. His immediate superiors and his doctors knew why he couldn't continue in the Marine Corps, or on any other job, but it still took that long to unravel the red tape.

It was financially necessary for me to continue working in order to keep and maintain our home and meet other than medical financial obligations. Tom could not go anywhere alone, for fear of being picked up by the police for public drunkenness. So while I was at work and the kids at school, Tom spent most of his life at home watching television, dependent on our return for outside contact. A very social being, Tom had many friends in and out of the Corps, but only a few ever came to see him after his illness was diagnosed, and only one came fairly consistently. Our couple friends also drifted away, seemingly unable or unwilling to cope with our new situation. Quite an adjustment for a strapping six-footer who always had had either a part-time job or involvement with scouting or Little League, or both, going along with his regular duties for the Corps.

As the disease progressed, Tom needed someone with him at all times. I was still working, the children were all still

in school and also working at part-time jobs. Household help, willing to take on the responsibility, was hard to find, and when found did not last long. Tom was not an easy person to be around. A month was our all-time high for any hired help.

He could not control his body movements and his moods were constantly changing. At a time in their lives when any teenager needs continuity and the stability of their parents to hold onto as they emerge into their individual adult lives, our children had a father who began turning on them one by one, and ordering them out of the house, then plead for them to come back. Only it never lasted for long and the cycle would repeat itself. I found myself in the position of secretly helping the children financially when they were ordered out, and reassuring them that we both still loved them and believed in them, that it was the disease making Tom act the way he did to them. A difficult distinction for anyone to make, let alone vulnerable teens. Whenever Tom found out or sensed that I was not in total agreement with him, I too became a target for his moods. It seemed to be very important to Tom to be calling all the shots. A once very proud, active, virile man who was used to having things go his way, having his every command obeyed without argument seemed to be the only vestige of manhood he had left.

Periodically, Tom would spend many weeks at the VA hospital in San Diego. He saw many neurologists with conflicting ideas on medication. All known medications for muscle and mood control were tried on Tom at various times. He didn't respond as others had on the same medications, and none proved effective for any length of time. They even fluctuated in their diagnostic, retesting, re-evaluating, trying to decide if he had H.D. or Oliva pontes cerebellum, a disease just as terminal, only with a more localized degeneration, longer life cycle and a better hereditary ratio, 1:12 instead of Huntington's 2:4. The neurologists wanted the psychiatrists to care for him; the psychiatrists said his disease was neurological. The end result was always the same. They would come back to Huntington's and the news that there is no known cure or help.

His hospital stays were getting closer together and our youngest child was almost 18 and already out on his own. I decided if I quit work and took care of him myself, he could still have a few happy times left. We could only manage to do this by selling our home and moving back to our hometown here in Colorado. This would also give the children a chance to make a life of their own without seeing their Dad daily progress downhill.

We made the move to Pueblo, where our many concerned relatives were more than willing to help. That was in March of 1975.

That first year he held out pretty well, even though the mood changes, fits of anger, and depression were getting worse. His speech was very difficult to understand. So slurred that sometimes he would have to repeat himself so many times, he would forget what he was saying. This inability to communicate with people frustrated him more than anything else. He would withdraw himself from one member of the family and then another; the slightest things would upset him. He developed a negative attitude toward everyone and everything.

Tom voluntarily was hospitalized briefly twice, once in the Denver VA Hospital and once in the Ft. Lyon VA Hospital. He underwent physicals, evaluations and changes in medication, trying to alleviate his moods and help stabilize his coordination.

As time passed, sleep became almost impossible for both of us. He'd roam the house day and night trying to find fault. Paranoia and distrust became a daily occurrence. He would follow me, never leaving my side or believe a word I said.

Our doctor in California had told me that as male Huntington's victims became more incapacitated they often felt the need to be exhibitionists, make lewd remarks, carry on flirtations, and were apt to accuse their mates of being unfaithful. Tom experienced all of these to some degree. He became obsessed with the idea of my unfaithfulness. No male in our narrow circle of contacts was free from suspicion. Tom and I will be married 25 years this coming May 9th and until this phase of the illness, he never questioned my loyalty or devotion.

Eventually he would begin feeling depressed over his actions, for which he had no control, yet realized they were not right, and he would apologize. His moods became more unpredictable and lasted for days at a time before he would snap out of them.

Our families became concerned for both of us, knowing that if Tom ever hurt me in one of his moods and later realized it, he would never get over it, and that I would find Tom's harming himself equally hard to live with. It was then that the suicidal tendencies became more noticeable. He would go for days

talking about doing away with himself. Whatever family members were in good standing at the time worked out a schedule and took turns, with some calling and others finding valid reasons to drop in daily. They could tell by my reactions and by looking at Tom what kind of day we were having. Sometimes their very presence would bring Tom back to a form of normalcy. We all knew we were just buying Tom a little more time at home.

Then on November 21, 1976, he tried to carry out his threats. I was able to disarm him, and one of our sons, who was visiting us, was able to subdue him by pinning him to the floor until one of my brothers and the police arrived. The police were kind to Tom, but because of regulations had to transport him to a local hospital in handcuffs.

That is when I had to do the hardest thing I've ever done in my life: take action to have Tom committed to the VA hospital at Ft. Lyon. I felt I was his last hope and link to the outside world, like I was playing God. And once I had made that decision, there would be no one to carry on and protect his hold on a normal life. I had prepared myself, in my mind, for Tom's physical deterioration to take place first, and was totally unprepared for him to deteriorate mentally first, unlike most Huntington's victims. Fort Lyons is a mental hospital where, in comparison to the other patients, he is at this time out of place.

With the help of the VA we are trying to have him placed in a nursing home in Colorado Springs so that I and other family members can visit him more often and try to reassure him we do care about his well-being and are trying to do all possible to keep him comfortable and happy. Fort Lyons is a three-hour round trip from Pueblo. I rarely make the trip alone. I never know what I'll find when I get there. Some visits are so peaceful, I wonder again if I committed him too soon. Others are so full of accusations from his obsessions and paranoia that I can barely endure them.

Our families have been more than understanding and try to help me accept the fact that Tom is better off in a controlled situation where he is not a threat to his own life. There is always that feeling of hopelessness, not knowing what he must be going through, and that there is nothing that I or anyone else really can do about it.

There are 29 patients at Fort Lyon in all stages and forms of this dreaded disease, beyond any means of help. When you see for yourself what the effects of this disease are, and realize how many at-risk people there are in our country, you want to cry out for help, to everyone to help, in whatever way he or she possibly can.

I know I have such a feeling of uselessness, seeing my husband deteriorate and the thought that in time, possibly in another few years, our children and grandchildren might be experiencing the same fate. So I am getting in the FIGHT against Huntington's and with the grace of God and the help of all good people, maybe we can help to stop H.D. before any more generations will have to face it.

Tom and I have been lucky in one respect, we do not have to face the financial burden because my husband is a veteran; but our children will not be so fortunate. Now that we know that we are a Huntington's family, our children are finding it hard to qualify for life insurance. Companies do not want to insure the at-risk group, or if they do, it's at a very high rate. So far they have not experienced health insurance problems because of group coverage at their places of employment.

Unlike many other terminal illnesses, it is possible for Huntington's families to live through the emotional stress, medical, financial, and legal problems not once, but as many as three times, and with more than one victim afflicted at the same time.

"There is a fate worse than death," and any Huntington's family well knows the true meaning of this expression.

TESTIMONY OF
DONALD ARMSTRONG, M.D.
ASSISTANT PROFESSOR OF NEUROLOGY
UNIVERSITY OF COLORADO

ARMSTRONG: My name is Donald Armstrong and I'm an Assistant Professor in the Department of Neurology and Neuroanatomy at the Medical School here in Denver. I'm also a consultant in neurochemistry at the Fort Lyon VA Hospital where there are approximately 25 patients now with Huntington's disease.

My research interest is in neurodegenerative diseases and we study these from a biochemical and pathological standpoint. Our research on HD has only really begun since the mid '60's. And only the creation--and only through the creation of local chapters--that is, local HD chapters--where private money is used to finance research, has any research really been possible. Research takes trained personnel. So, with limited funding, progress is very slow. Many investigators, I'm sure, are reluctant to begin projects in areas where money is not available, for the obvious reasons. Our lab, for instance, has been interested in determining whether a test could be developed for diagnostic purposes. That is, if we could get a test to work on patients that would identify patients with Huntington's disease, then we might also be able to use it for determining which of the offspring of these patients might be at risk. At the present time I receive a small amount of money from the Southern California Chapter of HD, and although we are making some progress, it is very frustrating to me not to be able to devote maximum effort because of this lack of funding.

Federal funding to study HD would certainly help us determine the exact biochemical lesion in Huntington's disease, develop a diagnostic test, and thereby offer genetic counselling. Money is also necessary to provide such things as a regional workshop where scientists can convene to discuss and share ideas in a real collaborative effort. I would also advocate the creation of a clinical research center at various locations around the country where patients could be admitted and studied by many health care professionals. We have such a center here at the University of Colorado, and myself and Dr. Neville have used this on many occasions and found it an extremely useful approach for studying other neurological diseases.

In summary, then, I see a critical need for financial support so that we in research can begin a sustained effort to study the causes of HD to be able to identify patients at risk, and thereby offer genetic counselling; and to investigate whether ultimate therapy is feasible. Certainly, what is learned from this form of presenile dementia will be valuable in our understanding of the other forms of early age-related changes of the central nervous system.

N. WEXLER: I would just like to correct for the record that your funding comes from the Hereditary Disease Foundation, and not the Southern California Chapter--it used to be.

ARMSTRONG: Yes, sorry about that.

N. WEXLER: Okay. I just wanted the record to be cleared. Your own research--you were originally involved on research on Batten's disease, isn't that correct?

ARMSTRONG: That's correct.

N. WEXLER: Is there a model, perhaps, for the way in which your research was expanded, which might be useful for other investigators?

ARMSTRONG: Well, there are some similarities in the pathology perhaps of these two diseases and so, researchers working on one area often can overlap into another in sort of a time-saving effort.

MCKHANN: Dr. Neville.

TESTIMONY OF
HANS E. NEVILLE, M.D.
ASSOCIATE PROFESSOR OF NEUROLOGY
UNIVERSITY OF COLORADO

NEVILLE: My name is Dr. H. E. Neville and I'm a neurologist on the full-time faculty at the University of Colorado Medical Center. I'm an Associate Professor of Neurology. I'd like to speak this afternoon from the standpoint of the clinician who sees an occasional patient--and I would say occasional--with Huntington's disease.

This is a rare disease in the experience of neurologists who, generally, at one time or another in the patient's history, will see the patient for diagnostic purposes. With over 25 neurologists in the city of Denver, this probably accounts in some part for the relatively low numbers of people, even at the medical center, that we see--the medical center, including the Denver General Hospital and the Veterans Hospital. This unfortunate disease, when the diagnosis is made, often results in great anxiety and depression on the part of a neurologist because he knows that there is no cure for this disorder.

There is no known enzyme defect. What is seen in patients who expire and in whom pathological examination is carried out is simply a lack of certain nerve cells and portions of the brain. And the reason for the absence of these cells, which we can only assume were there before, is unknown.

Huntington's disease is undoubtedly related to a number of other degenerative diseases which are more commonly known and more familiar

on the part of the general public. It is one of the degenerative diseases which includes Alzheimer's disease, or senile dementia, Parkinson's disease, and the somewhat rarer amyotrophic lateral sclerosis. All four of these diseases share the common pattern of different clinical symptomatology, but it shares the common pattern of being a degenerative process, which is a somewhat flamboyant way of saying that we don't really know what happens. We simply see the lack of certain critical nerve cells and we see the patient progressing through various stages of the clinical illness. When the diagnosis is made, there is no question that it is a tragedy for the patient, and a tragedy for all around the patient, both in the family and in the immediate circle of friends.

We must tell the patient at some time or other, and we must certainly tell the family from the beginning, that it is chronic, it is progressive, and it is invariably fatal. The diagnosis is based almost entirely on the clinical presentation of the patient, with some laboratory studies, which exclude certain other closely related but different disease entities. At present there is no laboratory test which makes the diagnosis with 100-percent certainty. Unfortunately, there is also no test to identify presumed carriers of the disorder--people who are quite normal in their teens and their 20's, but who may be expected to develop the disorder in their 30's and 40's. After the diagnosis has been made, the patient and the family are faced with the increasing problem of either mental failure, extraneous movements which cannot be controlled, even with a variety of new drug therapies, expensive home care, eventual nursing home placement, the possibility of being placed in a state institution, and all of this resulting in an enormous financial drain onto the family and friends. There is social deterioration within the family unit; there is divorce; there is suicide; there is a break-up of what might be a very closely knit family. The physician sees all of these things and is invariably unable to do the one thing which might remedy the situation in a more meaningful manner; and that is, he is unable to treat and he is unable to cure.

Now, with this gloomy picture, which I am sure the Committee has heard time and time again, the question is, "What is needed?" In my view, at this particular time, when our knowledge of this disorder is so rudimentary, we need most desperately a method of identifying patients who will develop the disease--that is, family members who are currently unaffected, who can be identified with 100-percent certainty and can be counselled that if they are to marry and have progeny, the chances are one out of two that they will have affected children. I think this is the only way, at the present time, that we can in any way control the continuation of this disease in our population.

Secondly, we need adequate research funding to support research measures to identify the cause of this disorder. And it is my view

that the cause of this disorder may well be found in our lifetime and may give us clues, in addition, to the cause of these other degenerative diseases, including amyotrophic lateral sclerosis, Parkinsonism, and Alzheimer's disease.

Finally, I support any and all research efforts which will lead ultimately to adequate treatment for this disorder and a cure. However, I think that an enormous amount of research is still necessary to develop a cure because we still are uncertain as to the cause of the disorder and this is something that to me is a great distance away. At present, I think the development of a diagnostic test for this disorder would be of the highest priority in all of the priorities that I have mentioned.

Thank you.

MCKHANN: I would just like to thank people for coming. I have to leave if I'm going to get back to Baltimore in time to function tomorrow, and I think this has been very helpful. We have some more testimony to take. Marjorie Guthrie will be right back, and I'll ask Dr. Wexler to take over the microphone.

N. WEXLER: Part of the problem is that the only flights which go back East leave, I think, at 4:30 or 5:00, or else you have to stay over. So just remember that you're speaking, not to individuals, but to the Congress and President.

I had one question either to Dr. Armstrong or Dr. Neville. We have gotten many recommendations that there be some kind of a clinical center that would combine care for patients with clinical research and, of course, we can't put centers wherever we have heard these recommendations. Since you're so close to the VA, and have VA connections, would it be advantageous do you think to have some kind of center through the VA that would have some kind of special program for perhaps neurological patients, or dementia, or presenile dementia patients, or some catchment of patients, so that you would have access to a research population as well as providing better care for these patients? Is this something that has been even approached with the VA?

NEVILLE: I'm not aware that it's been approached in such a way with the VA, but one of the problems is that in approaching the VA about projects that would involve medical schools or medical centers, there's a great deal of reluctance to become involved. And a good example is the fact that the VA would be most unwilling to share, in any way, a particular bit of instrumentation like the CATscan. Now, this is an extraneous issue, but it's a good example of the problems that we have with the VA Administration.

N. WEXLER: What on earth was the rationale?

NEVILLE: I have no idea.

N. WEXLER: But you should go out and buy your own, is that...

NEVILLE: That was a possibility until we considered the cost.

N. WEXLER: That's right.

NEVILLE: I think what I'm saying is that the Veterans Administration--and I'm talking about the central office in Washington--has to give backing to local initiative in developing programs that you're discussing. I think unless this backing comes from the VA central office there's no chance of getting funding, though there's a lot of lip service paid to cooperative projects and, indeed, there is a dean's committee which works very closely with the VA Administration locally. Nevertheless, when one talks about any sort of cooperative effort, one faces the possibility of considerable red tape before the subject is even broached to VA central office. And it's been a bit of a discouragement to some of us who have tried to develop programs in the past.

GUTHRIE: Perhaps, now that we've shown how many patients are in the VA with HD, maybe we can pressure a little bit of cooperation since it's costing the VA so much money for the care.

NEVILLE: In custodial care.

GUTHRIE: That's right.

NEVILLE: Yes, I agree with you completely.

N. WEXLER: If you have certain specific programs that you have been discussing with the VA, it would be helpful to us to have those specifics since we are looking--in fact, have a group of people who are visiting with the VA. They'll be testifying before the Commission in May. And so, if we have access to what kinds of programs you'd recommend there, that would be very useful to us.

GUTHRIE: Thank you.

Did someone explain why people disappeared? They had to go back to their various places, and I want you to know that Dr. Tower, who is a director of the Neurological Institute, also had to leave. But it was wonderful that he stayed most of the day. He was able to hear them. He said to me as he left, "I've learned a lot."

TESTIMONY OF
JOHN ARCOTTA
BOULDER, COLORADO

ARCOTTA: My name is John Arcotta, I reside in Boulder, Colorado. I am 35 years old and "at risk", I am only now beginning to piece together my background and learn about Huntington's disease. There is some discrepancy about when my father actually died. A hospital report says 1942, a newspaper record and funeral card say 1944, one of my older brothers told me 1941, three months after I was born. To my knowledge, he was not a victim of HD. After my father's death (I never knew him) I do not know what became of my mother. State hospital records show that she was committed in 1951 and died of HD in 1956. I do not know where she was from 1941-1951. I do not know who committed her to the state mental hospital. I never knew my mother. I did not know she was alive from 1941-1956 until I was notified of her death in 1956. I attended the funeral. That was the only time I saw my mother. It was a bewildering experience for me.

After my father died, eight of us children were living without parental care in a project in Bridgeport, Connecticut. Through neighbors, the state welfare was notified of our plight and put us temporarily into an orphanage. My oldest brother, Blazie, who was around 16 then, did not go into the home but tried to make it on his own. I am sure it was a tough experience for him. He saw the family split apart and he was unable to do anything about it. He did not know what was wrong with our mother. The schooling of the older children was immediately discontinued as they tried to find jobs and support themselves and us the best they could. I was placed in several foster homes. Mistreatment by foster parents kept me moving from one home to another. I was confused, lonely, unhappy, and constantly in search of "my family" that I heard, now and then, existed somewhere. I quit school after the eighth grade. No one really cared. I went to live with an aunt and my brother Larry. This was a very good period in my life. I was very close to my brother Larry. He was committed to the state hospital while I was in the Army. He was admitted in 1961 with a nervous condition. He was diagnosed with HD shortly after and died of HD at the age of 38 in 1976. I was always told by other members of the family to not go see Larry. They told me it would horrify me. I have always regretted that I did not visit him. Now that I know more about HD I feel very sad that I did not visit him.

As we were growing up, all of us felt that our family suffered from hereditary insanity and that we were singled out as a family with this "crazy streak". We were taunted with "Your mother is in the crazy house!" I did not then even know that my mother was alive, and if she was alive and in the crazy house, I did not know why.

My mother, two uncles, two brothers and one sister have been diagnosed with HD. My brothers' and sister's spouses all divorced them immediately after committing them to the state mental hospital. They refrain their children from visiting with them and have completely abandoned them to die alone. My brother, Blazie, diagnosed with HD seven years ago, has not had a visitor (former friends or relatives) in many years. He helps out at the hospital. He is still active and alert. However, he is frustrated and cannot understand why all former ties were severed so abruptly. My wife and I have decided to have him come live in Boulder with us as soon as we can make the proper arrangements and transfer him here from Connecticut. We will do all we can to make his last years as good as possible. He will not be left alone any longer.

I have no formal education to speak of, no training in a skill other than music, which is a major part of me and has sustained me for many years. I have developed my own business in recording since I got married nine years ago. My wife knew I was at risk when she married me. We have two beautiful sons. My wife's parents know that I and our two sons are at risk. Their attitude is very optimistic and supportive.

They feel, as we do, that if there is a 50-percent chance of inheriting HD, then there is also a 50-percent chance of NOT inheriting HD. They and we have chosen to think positive and plan our lives in a positive direction. Of course, we admittedly have those moments when a stumble or dropping of something triggers that thought, "Is this the first sign of HD?" When I become depressed about these things, my wife assures me that she is pretty clumsy herself and so are a lot of other people. She has also assured me that she will care for me and will not abandon me if and when I develop HD. With my background it was difficult to believe that she meant it, but I have come to know that she is sincere. We both have become active in the Rocky Mountain CCHD Chapter, and despite many setbacks and much discouragement we will continue to strive toward a better understanding and treatment of this disease.

I would like to say here that at-risk children should not shy away from marriage, having children, and leading a normal life. You must take a chance on life. You must develop a positive attitude. HD is not the only disease in the world that kills. Other people suffer loneliness, child abuse, cancer, murder, accidents, wars, insanity, MS, other genetic disorders, et cetera. Life continues. You can either be defeated by the challenge, which means neurosis. You can conquer the challenge, which means growth. There are no other choices. Being an HD victim is not any fun. Caring for an HD victim is not an easy task. Being at risk can be a burden. I have seen my family split apart generation after generation because of ignorance, hopelessness, circumstances, confusion, weakness, fear, inability to deal with "the facts." I want to change this. With the help of the Commission, perhaps we can all help each other. Following are some suggestions:

We must become educated, first of all, before we can understand and realize that HD need not lead to separation, divorce, suicide and despair. With knowledge, support and compassion we can continue to care for those we love afflicted with HD. We need to educate our doctors and the public of what HD is. We need places to go for help. We need nursing facilities where HD victims can be cared for--they do not belong in mental institutions. We need people who understand problems that HD victims and families have and how to deal with these problems so that they may have more productive years than they would have without any support.

We need the following:

1. A national campaign effort is feasible and should be initiated by our National CCHD office, which has the connections to organize and promote such a campaign. Benefit concerts with top entertainers, TV spots, candy, cookie or card sales, telethons, et cetera. Jerry Lewis has done well with muscular dystrophy. Perhaps we could find someone with such dedication and interest to develop a campaign with equal success. Local chapters can only do so much. We need National support. HD needs to be brought to the attention of the public. I believe that National CCHD staff members do have connections with the movie industry, with the entertainment business, they are well-travelled and involved with influential people. Contacts could be made. The opportunities are there.

It is common knowledge that genetics is being brought more and more to the public. Research is advancing rapidly. Our children are learning about genetics in school.

2. Regional Centers where HD patients and families may write and/or visit for information and assistance. These centers could be beneficial to those families already inflicted with HD as research continues to seek our cause, control and eventually a cure of this disease.

Each center (ideally, one per state) should offer the following:

1. Diagnostic Center
2. Drug Center (medication)
3. Care Center Referral Service
4. Genetic Counselor
5. Social Worker
6. Legal Service
7. Therapy Center (physical, occupational and vocational)
where patient and family could be instructed on care to keep HD patient independent as long as possible--to keep patient a part of the family unit

8. Financial Aid
9. Insurance
10. Volunteers--to aid families in caring for the HD patient. These volunteers would be specifically trained to care for HD patients. They could assist the families in home care, shopping, staying with the patient for short periods of time so that the family can maintain a normal life pattern with less strain.

Of course, I realize that the above suggestion would take time to set up and funds to support. It is a goal to strive toward, however. Meanwhile, perhaps the following options would be more easily established:

A staff of two or three individuals who would be knowledgeable in the facts of HD and the needs of its victims. These people would make contacts within the state with doctors, social workers, lawyers, et cetera. HD families could call, toll free, and get information they need and be referred to the proper person for assistance. We cannot educate everyone at once, but a few could be educated at a time and these people in turn can help those who need help. It is discouraging to go to a doctor who knows nothing at all about HD. HD patients should know which doctor they can go to for care.

The second option I have to offer should be established within the year. A nationally distributed booklet must be compiled which would list the following:

1. Local CCHD chapters and other related organizations
2. Health care centers that are capable of diagnosing HD and giving medical treatment and advice
3. Problems that HD patients and families encounter and services that they can refer to for assistance. List addresses and phone numbers.
4. An updated section on the care of the HD patient.
 - a. Description of HD symptoms and what to expect
 - b. Care Plan: Good hygiene-maintenance of elimination, muscle use, crafts, emotional needs, nutrition, et cetera
 - c. Helpful hints for HD patient to aid him in remaining as independent as possible.

This information can be compiled from people who have worked directly with HD patients, either professionally or family members who know by experience.

These centers can be staffed economically if properly trained individuals are employed. It is possible to educate the layman, the volunteers, members of HD families, relatives, caring individuals.

Those people who have been a part of, or witnessed a friend with HD are the best qualified to know and understand the mental strife that occurs. With proper training (brief and effective) many individuals can be of service in these centers. We do not need specialists with degrees--we need people who know and care.

HD families need to know that there is a place close by or at least a book where their questions can be answered, where they can receive support and assistance. (I feel we could get Federal funds to publish the booklet I suggested be published.)

More often than not, the HD patient is left alone, confused, lonely, helpless. At-risk family members suffer depression and fear. We must assure these people that they are not alone, that there is hope, that they are still capable of loving and being loved, that their lives can continue.

Of course, a cure for HD is the ultimate goal, but we must not overlook, in our striving for this goal, that there are many who now suffer and will suffer before a cure is found; we must not forget them, and we must now, as soon as possible, with as little red tape and long-winded reports, excuses and ignorance, set up centers of care and treatment and/or publish an informational and instructive booklet on the care of the HD victim.

Thank you.

GUTHRIE: Thanks, John. I think that all the recommendations you made are things that this Commission wishes to provide. And that is the purpose of this hearing--to be able to get what we need. However, you know that until this Commission submits the final report, and until we organize and fight for the funds to implement this report--even if it's accepted--it is going to take quite a bit of time. And I would remind you, therefore, how valuable it will be for all of our families to help us in doing something, even now, towards the goals that you so beautifully stated.

And I want to tell you that, for instance, just two weeks ago, we received at CCHD a beautiful, up-to-date mini-review that I would love to see circulate to every single doctor and every single family on our mailing list. But it will not be done until we can raise the money to do it. Money is, of course, what holds what we can do, until the Commission can make better and more feasible answers to all the things you've said.

The other thing I must comment--because you brought it up, and I want to say publicly--yes, we have connections but we cannot do a Jerry Lewis telethon--not because we don't have the friends; it's because if you would look at it...did you know that muscular dystrophy

as an organization has 5,000 employees, to say nothing of the volunteers. The other agencies have 2,000 and 3,000 employees, to say nothing of the volunteers. The Committee to Combat Huntington's Disease today has 5 employees; I am a volunteer. And the next agency closest to us is the autistic children, who have 13 employees. So it isn't a question of just getting the people in the public eye to help us; you must also have a staff, and that may take a bit of doing. But we're doing what we can, and with your good help I know we will do better.

Do you want to comment?

ARCOTTA: Yes, I've tried in the past to promote three concerts.

GUTHRIE: How well I know.

ARCOTTA: And one of them, in June, was very costly; and for the time that it took to organize the concert, which was approximately three to four months, I think the Rocky Mountain Chapter raised something like, maybe \$200.

GUTHRIE: It's unfortunate, isn't it, that our membership didn't involve themselves more, but we have to educate there too.

N. WEXLER: Could I say something for a moment, because I think it's important, especially since this testimony is transcribed in part of our Commission's records--that there are a number of different groups that are involved in Huntington's work, and I think all of them--or many of the people here--are part of the local chapter of the Committee to Combat Huntington's Disease, which has been a major force in organizing the HD community.

As part of our role as a Commission--which is another hat that Marjorie wears--we are to make recommendations about the role of the private sector in HD support; so that partly what we do as a Commission is that we will make to the government, to the public, to CCHD, recommendations about their roles, the ways in which they can be useful, and what they need to be useful. At the moment there are four different health voluntary organizations dealing with Huntington's--CCHD, the Hereditary Disease Foundation, Huntington's Chorea Foundation, National Huntington's Disease Association--so that there are a number of different organizations just for HD in the private sector about which we as a Commission can make recommendations, and you, as a public, can make recommendations to us.

Now, if I were to translate some of your suggestions, it sounds as if the health voluntary groups can be tremendously effective in organizing families and even providing some services for families. One of the recommendations may be that we need to

recommend, perhaps, that they get some Federal support in order to carry out some of their programs; that they would need Federal support to put together this kind of informational booklet, for example. But I just wanted to clarify because I think that people have been, you know, using various names for the Committee--the Commission, the this and that and the other, and there, the council that at least this body is not part of any private group and can make recommendations about all private groups.

ARCOTTA: What if this fails? I mean, I hate to look at it in that light, but we also have to take into consideration there is a possibility, if it does. Now, I might also add, I've been in music for 20 years. I started playing professionally when I was 14. I was in the musicians' union for 20 years. The way I feel, a fund-raising thing is to get--just get some good concerts all over the country. And I think it can be done with very little red tape; I just honestly feel that it can be put across. Well, when I see a lot of concerts and benefits happen that are successful--for instance, I know of a piano player here in Denver--somebody stole his piano and his clothes. Okay. The piano may cost \$1,000; maybe his wardrobe was worth \$500--and they had a benefit concert for him. They raised \$5,000.

N. WEXLER: I think people can identify with lost clothes more easily. I think that you raise an important question, I think, in some ways, it might be more relevant for the chapter. But what I would suggest is that--I think we have one more group, and maybe after we hear this group, then we can open up for all kinds of questions.

ARCOTTA: Okay.

N. WEXLER: Thank you.

Mr. Larry Boyd, Robert Christ and Jill Heesan, please.

TESTIMONY OF
LARRY BOYD

BOYD: First, let me start off by saying that I did not submit a written testimony and so I'm, therefore--I came in as an observer.

My mother is a victim of HD--and not having any prepared testimony, and hearing a lot of what's been said already--there's not much that I think I can add, other than to say that I'm very heartened to be able to come and hear. It's been an uplift for me just to hear some of the other problems that the other families, and whatnot, are going through. Many times, I think, with the isolation of HD victims we tend to feel that we're alone in terms of our situations and experiences with the HD victim. And it's been very

uplifting for me today to hear the different situations that relate directly to my experiences. It kind of takes the weight off of my shoulders to know that other people are going through the same kind of problems, because I think many times we sit back and in our loneliness feel that we are the only ones experiencing a given situation.

My mother has had HD for the last eight years and I am one of six children. We are all aware that we are at risk. We have two brothers who are doctors in California, and through that, we have been able to...with medical people in the family, I think, it's been a lot easier for the six children to understand and accept what is going on. From that standpoint, many of the things that I've heard today have been so uplifting to me, because our experience has been mild compared to some of the things that I've heard testified to here today.

I would like to say that there is a tremendous need in our particular situation for in-home care. One of the major things that we found has been a block to this--keeping an HD patient in the home--we find that they are owners of property and they have assets that many times may take them \$2 or \$3 over the stated limits of the law. And as a result, we have not been able to get visiting nurse service in the home, because of the value of the home.

We've gone through the adjudication process--that was thrown down--stricken in terms of the state law and a new adjudication process has now come into being. As a result of the first filing for adjudication, the six children sustained a debt of about \$1,500 to \$1,600 which we split between the six of us, and have just completed paying. My mother has been restored to the home, since they struck down the old statute, and she is now in a five-bedroom home by herself, and with very little care except for what we can do in terms of getting by to see her around our jobs and our own families. So from that standpoint I would like to see if there can be some things done in terms of the improvement of the ability to take care of the HD patient in the home. It's been mentioned many times today that that's a goal that I know the Committee is shooting for, but I would like to re-emphasize from my experience. It's very difficult to keep an HD patient in the home and get around the governmental regulations--the governmental guidelines--the governmental restrictions, in terms of assets and income that a person can have.

We have also been through the genetic counselling over at the University of Colorado and have talked with a number of the scientists and doctors who are working in that area. And so, from that standpoint, we're very knowledgeable of what's going on and very grateful to what's going on over at the University of Colorado and through the National Committee. We've also been helped very greatly by Mr. Seward Wilson, the local chapter president, and he's been very helpful to our family, personally, in getting through the problem.

But I would just like to emphasize that we do need help in the areas of keeping the victim in the home and being able to provide services and, you know, get people who can come by and see them and that kind of thing, and get around a lot of the governmental regulations that prevent--many times almost force--you to put the person in a home. And I would like to see the research continued and hopefully some things will come out of that that will help us all.

GUTHRIE: I call that the middle-class dilemma. Either you're too rich or you're too poor.

BOYD: Precisely.

GUTHRIE: But the tough one is that one in the middle-class dilemma, and we do have to work on that; yes, indeed.

BOYD: That's all I have.

N. WEXLER: I don't know if you were here before, Mr. Boyd, when Dr. Anna Marie Lubs was testifying about a program that she wants to start of group counselling, long-term group counselling with HD families and other families with genetic diseases. Were you here then?

BOYD: No. I think that was during the morning. I just arrived this afternoon.

N. WEXLER: If I didn't do violence to the research proposal, it was to have ongoing groups over a six-month or a year period. Is this something that you'd be interested in doing for yourself, or do you think that any of your brothers and sisters would be interested in this?

BOYD: Yes, I did hear a comment, I guess, from the lady who had children, been in the research studies, and whatnot. My oldest sister is 33 and we are all kind of sitting on pins and needles now hoping that she'll get by the--you know, get through the period of the major or the general age that it takes onset. So, we're all kind of sweating, but the fact that we are all a lot older and the period through the genetic counselling hasn't embittered us to the point that we are not willing to participate in any other studies. What we found is that through eight years of going through things and not having the support and whatnot, that you just give up and you just hope. We sit back each day and hope that she doesn't fall in the home or that something serious doesn't happen.

But we're more than willing to participate in something that seems worthwhile. We've been through so much that has been kicked back in our faces, or has been turned around by some small legal

regulation or something like that, that we've given up in terms of searching out the social service agencies and that kind of thing. But in terms of participating in studies and that kind of thing, we're open to it.

N. WEXLER: Well, this is a research project, but the research question is, is this valuable for people who are at risk or where spouses have the genetic disease in the family? And so what we would want to know is--the Commission making recommendations about kinds of research to fund, kinds of programs to fund--would you, as a person at risk, or do you think your brothers and sisters, would find it useful to not just go for one- or two-shot genetic counseling but something that was ongoing that would be a group of your peers?

BOYD: Yes, I would say. From my own personal standpoint, I've just kind of reached the point that being an at-risk person doesn't pose any problem for me. I'm going to go ahead and live my life. I've just personally reached that decision within myself and most of my brothers and sisters have also. So, from that standpoint, we don't feel that nothing would be worthwhile, but I'd be willing to participate.

The testimony of the young man was quite moving to me because that was a big uplift, specifically, because he was my age; and on the other hand, has not been able to develop the attitude of, you know, "I'm going to go ahead and do what I need to do." So, from that standpoint, it was nice to hear that there was someone who has different feelings. But I would be willing to participate in something like that.

N. WEXLER: And I think it kind of uplifted him when you came back and said what you did.

BOYD: Oh, well, good.

GUTHRIE: Sharing means a great deal to all of us, doesn't it?

BOYD: True, it does.

N. WEXLER: Robert Christ.

TESTIMONY OF
ROBERT CHRIST
ASSISTANT DIRECTOR
MEALS ON WHEELS

CHRIST: I'm Bob Christ from Metro Meals on Wheels. We provide a service that some of the families may be interested in. We deliver

hot meals during the noon time, five days a week, or on weekends, if they need it, at a cost of \$2 a meal. You get a choice of two entrees a day--for example, today we had French dip and tuna casserole. We deliver them in a reheatable container--it's aluminum. And this service is provided to just about anyone who wants it. Mainly, we service senior citizens, invalids, or just anyone who is incapable of cooking their own meals.

GUTHRIE: And how do people get to know about this?

CHRIST: Okay. We are listed in the phone book under Metro Meals On Wheels, or I could give the phone number where we could be reached if anybody needed it. We can be reached at 255-3161.

GUTHRIE: Are you privately endowed, privately funded?

CHRIST: We are part of ARA services, which is active in a Meals On Wheels Program throughout the country.

GUTHRIE: All right, tell us, what is ARA?

CHRIST: At one time it meant Automated Retailers of America. We are a private company.

GUTHRIE: You are a private company who provides service. It's a very small fee, I gather. And they go anywhere in the city?

CHRIST: Within reason. We service about a 100-square-mile area at present.

GUTHRIE: I think that's very helpful to know.

WEXLER: And there's no eligibility requirements?

CHRIST: No, all you have to do is give us a call--a day's notice--and we can start delivering. We do not deliver on Saturdays and Sundays, though. But people who participate in the program can have extra meals delivered to them on Friday and all they have to do is put it in the oven and heat it up.

GUTHRIE: Very good.

N. WEXLER: Do you know if you service any Huntington's families?

CHRIST: Not to my knowledge.

GUTHRIE: I'm sure that is a great help to someone who wants to live alone and be independent and doesn't want to take the trouble, or cannot take the trouble, to provide her own meals. That's an excellent--it should be funded some day. That might be a very good suggestion for the Commission to recommend this kind of funding.

CHRIST: There is a bill before the Senate called Title VII that would provide funding for us to run this on a much larger scale. Right now, we're running this out of another unit using the people in the other unit to provide the cooking and the staff necessary.

N. WEXLER: You're talking about at the Federal level?

GUTHRIE: Yes, I'm assuming--you're saying Federal level, but I suppose it could also be state level,

N. WEXLER: Is your company nationwide?

CHRIST: Yes, we are. We're the largest food service company in the world.

N. WEXLER: And is your funding totally private now?

CHRIST: Yes.

GUTHRIE: Could you submit something--whatever you use to publicize--to the Commission so we might have this to tell people about it in other states?

CHRIST: Sure.

N. WEXLER: Do you know what the Title VII is of?

CHRIST: I had the bill's exact number, but I can't find them right at this time.

N. WEXLER: Okay. If you find them, write them in.

GUTHRIE: Yes, we should get it. Ms. Heesan.

TESTIMONY OF
JILL HEESAN
AURORA, COLORADO

HEESAN: My name is Jill Heesan and I'm a member of the Rocky Mountain Chapter of the Committee to Combat Huntington's Disease. I'm going to make my remarks brief this afternoon since everything and every situation has been talked about. I could repeat, but I won't do it since you've already heard it and you all know the stories and the horrors as they were told.

My mother is now dying of Huntington's disease. She was declared mentally incompetent as a paranoid schizophrenic and was committed to the State Hospital, where shock treatments were administered repeatedly. My beautiful mother never recovered from the

great pain and suffering which resulted from this dreadful misdiagnosis. The disease has progressed until now she is completely helpless, weighing only 57 pounds, unable to talk or sit up, just lying there in a fetal position in her bed with only her eyes still alert.

We first heard of the disease in 1956 when we first saw our aunt, who subsequently died of HD, and learned that our grandfather possibly had the same thing, since he died with a "shaking palsy". However, when our mother became ill, we were assured that she exhibited no symptoms of HD, even when we told the doctors of our family history, as we knew it. Not too much later, we learned that our uncle had died with the same symptoms.

It wasn't until 1967 that mother's HD diagnosis was confirmed, when we learned the hereditary implications of the disease. By that time, each of us, my brother, two sisters and myself, had married. We all had children, and I had two grandsons. We had a conference with two doctors, my sisters, myself, and our husbands present, and were told that HD was a rare hereditary disease for which there is no cure, and that conclusive proof of HD can only be obtained in a postmortem autopsy.

Even though Mother had lived with my sisters and their husbands during a portion of those years, upon learning of HD, their marriages became intolerable. Both were deserted and each was left with three young children to raise. None of us, as far as we knew, had inherited the gene, but how could we know when there are no diagnostic tools?

One of my lovely sisters died at the age of 35, not necessarily because she had HD, but because she was afraid and an uncaring professional individual told her "there was nothing wrong--she was just wasting their time." I am now the legal guardian of her three sons, whose father hasn't even written them a postcard, even when he was advised of her death.

My brother, sister and I have all had problems with deep depression, and had neurological tests which have been negative so far. Is our depression a symptom of the disease, or just anxiety? We will never be certain that we aren't HD carriers until a sure method of diagnosis is developed, without which there can be no cure. This can only be accomplished through research. We, all of our children and grandchildren are at risk.

I have been a member of the Rocky Mountain Chapter of the Committee to Combat Huntington's Disease since its inception in 1970. Our organization strives to advise HD families and assist them, educate the public, and raise money for research through volunteer efforts. It is apparent that we alone are inadequate. I urge the Commissioners to report back to the Congress the immediate need of funds for research, family assistance and clinics for HD patients.

GUTHRIE: I don't have any questions to ask you except to tell you how happy I am that you mention the fact that we can do only so much, and that the real effort will come when everyone goes for the millions, not the thousands. The millions need the thousands to promote it to get the millions, and so the millions are going to come from our Federal government when we get the thousands to promote it.

N. WEXLER: I guess I would like to ask you the same question that I asked Mr. Boyd. Is there anything that, for you and for your family right now, that could make your life more comfortable in dealing with the anxiety of being at risk?

HEESAN: We did not find counselling to be successful, as such. Both for my sister, who is dead, and my other sister Shirley, who is just sort of floundering through counselling at this point in time. And I myself thought I had found a young man who was going to work out real well and I could work out some of the problems. But then he advised me in a moment of confidence that he felt rather strange around those of us who were HD patients and those who were at risk. He sometimes got the feeling that it might be catching. So we have not gone back.

GUTHRIE: I'm glad you said that out loud so that we can do something about that, perhaps.

WEXLER: I hesitate to ask this person's profession, but what was he, do you know?

HEESAN: Yes, I do; he was a psychiatrist.

WEXLER: Psychiatrist? Do you think that if you found a person who had some training and who had some experience with genetic diseases and Huntington's disease, that counselling is something that you might look to again?

HEESAN: Yes, I believe so, because we've met people like Dr. Kimberling and Dr. Armstrong--wonderful people down at the University of Colorado who really have a heart and understand--are trying to help us. Now, these are the type of people that can provide the assistance that we all need.

BOYD: I just wanted to say that I think in our struggle to resolve this problem of HD, many times the at-risk people and the people who are involved with HD do not take it upon themselves to make sure that they scrutinize medical personnel. I think it might be well wise for all of us to get away from the syndrome of walking into a doctor's office and believing that because he's a doctor he's competent. I think we need to test people out before just blindly laying out what's going on, find exactly what they know

about the disease, because I'm quite sure that we all know what the disease is and what it is doing and what the implications of it are. But we too often accept the fact that because a doctor is a physician, he knows all about all diseases. We need to scrutinize ourselves and make the medical profession become aware of what HD is; refuse to go to the doctor if he does not show any knowledge, or at least encourage him to get in contact with Dr. Kimberling or some of the other physicians that we know to be competent in the field; and make sure that they get some kind of training.

GUTHRIE: It might be interesting for you all to know that in any textbook of neurology, up until 1972, the only thing a neurologist might have seen or heard about Huntington's was a paragraph about this big. So, you must remember that the first textbook ever published on this disease came out, actually, in 1973 as a result of the Centennial Symposium on Huntington's disease which was held in Columbus, Ohio in 1972. So we cannot just tell the physician that he doesn't know his business. There was no place where he could get the information. Fortunately, now we can say, "Doctor, you can be educated if you would like to be." There is basic important information about this disease.

N. WEXLER: But I think that the point that he raises is an excellent one and I think it's a general one that people tend to assume, because somebody has a white coat or a degree that they know what they're doing, and I think that that's not true.

As a psychologist, I would advise any person that I saw professionally, if whomever they're talking to was saying something that's nonsensical, then question that person and walk out and find somebody else. Do not stick with somebody who clearly knows less than you and is being destructive.

Just one other question for both of you. Have you ever been, either through the Chapter or on your own, part of any kind of a rap session--a group with other people who are at risk?

HEESAN: Our own chapter, most of the time, is a rap session because new people who come are just like we were when we first became members, not knowing anybody else even had the disease.

BOYD: There are some stigmas involved and I think--for my brothers and sisters and I--we've all gotten past that. But there are stigmas involved, I think, that are attached, that prevent one from wanting to let other people know that you have this problem or you have someone in your family who has that problem. And I think that's something that we all personally have to get over, but that many times prevents rap sessions. But I think the Chapter is the one place that you can find a place to talk with people about the disease.

WEXLER: I would say that in my own experience of being at risk that the most helpful people to me were other people who were in my same shoes. And one thing that you can do, even before the Commission makes its report--before we get funding--is to get yourselves together and see if you can't find some knowledgeable person who would come participate with the group. If you were a group, you can even hire somebody, if need be, but you probably could get some professional to volunteer. And just from the kinds of things that I've heard today, it sounds like everybody is so isolated and separated and apart that between now and September just get yourselves together and talk for an evening. I think you can make your own beginning, and you probably are the best therapists for yourselves than any other professional person.

GUTHRIE: But you also have wonderful professional people in this community who are willing to come if you want to lead the group, which I think sometimes is needed, too. This has been our experience. I will tell you of a story of a group of young people, a physician of 24 who was at risk. He was the first young person I had met who was a professional person and who was at risk. So he was willing and we sent out word that if any young people between the ages of 18 and 24 would like to talk, they were welcome. Twelve people came together from Massachusetts, Philadelphia and New York, and I think one from Maryland. And they all sat down together with this one professional person who was at risk. And they had a very interesting discussion. The report came back, and we asked them, "Would you like to have another one and perhaps bring in another professional?" They said, "No, we would just like to talk again. We have enough of a professional. We've got our one good doctor there at risk and he was great." And it was a very helpful discussion. I think we've come to the end of the formal part of our meeting, however, the floor is open. Is there anyone else here who would like to testify?

I know Lois asked if she could have a few more minutes and you are most welcome now. Lois Reed, won't you come forward. And is there anybody else who would like to come forward? Please do come forward. That's fine, we have our three chairs filled.

REED: The only reason I asked for a couple more minutes was because I was very nice and kept mine down to five minutes, I think. In listening here this afternoon, I'm not sure but I had an idea of something that might be a recommendation. I, just a few minutes ago, talked with a young lady that had been sitting near me who has recently been diagnosed as having Huntington's disease. She was unaware that there were other people her age, and so on.

All right, here's my thought. I get letters and phone calls, from not just in Arizona, but all over, saying, "Lois, just stay there. We've got to know that you're there." I think our Chapter as well

as the other chapters of the Committee to Combat Huntington's Disease are vital. I don't have the heart to tell these people that I can't stay there too much longer, for a lot of reasons. But I think we have to keep our chapters of the Committee. I think in counselling, many times people getting together in the same boat can be as effective as professional counselling. I can't find anyone to take over my job. There seems to be no one else who is willing to give their home as an office, which we've done because we couldn't afford an office, and serve seven days a week. The people who are interested can't do it, they have family members who are afflicted, I don't. My thought is, would it be possible to recommend to the Commission that there be some sort of funding, instead of creating something new, I mean in one area--I know this is just a small area--possibly funding for some of our local chapters or our National Committee to Combat Huntington's Disease? Several people there in our area have said, "What we have to do is get an office and hire one person full time to do what you've been doing for nothing." That would take just about what our local contributions are each year. We'd have nothing left to fund our local clinics, and so on. Would it be too unrealistic to suggest that you recommend to the Federal government that there be some kind of funding for an organization that could do roughly what our Committee to Combat Huntington's Disease has been doing? That's all I wanted to suggest.

WEXLER: It is possible and I think that there are precedents in other groups. The way that it would work is that the organization would have a specific project and it can work two ways--either you can apply to the government and the right department in the government for a grant for that project, or the government can contract with your organization to do a particular piece of work if you can demonstrate that your organization is the best possible organization for the job.

And the government, being the government, generally has allowed different groups to compete for funds. So since there are a number of different HD groups, as well as a number of other private and public groups which could possibly do a job, then they would compete for a certain grant. But it certainly is possible that something that the Commission is considering and it means that you need to provide us with specifics of the kinds of things that you do do, and your qualifications for the job that you're manning.

REED: Thank you.

KNISS: My name is Juanita Kniss and I'm a teacher at South High School here in Denver. The reason I'm here today is because my mother had Huntington's disease and my daughter who was here with me earlier and I had gone through much of the hell that had been mentioned by the other people. But this morning, in listening to a lot of the men scientists talk, they talked about high school education only in terms of kids learning through science courses, and I teach social problems. There are courses in marriage, in the family,

sociology courses, just a wealth of outlets for this kind of information. I was thinking in social problems, which sort of is like sex education and family life cycle, we talk about family planning. And I think family planning centers would be great places to get a hold of young people to tell them all about this problem. As rare as the disease is, I've had three students from South, coming from a family of six, whose father has it. I've learned so much today that I can help that youngest girl with and I just want to thank you all.

WEXLER: Thank you. I think that's an excellent suggestion. Nobody has made it before and I'm delighted that you came and testified. And, in fact, you strengthen one thought we had, which is to try to grab everyone at Planned Parenthood and get them going. Thank you.

GRIFFIN: My name is Pat Griffin and my father had this disease. My brother died from it too. My brother died in '73 and my dad died in '62. Well, I was told last summer from my doctor, who is a neurologist, Dr. Earnest, that I had it too. And so, I'm under his care now and I have to be. But I live alone and I can for now, he says. I'm on medication and I fall once in awhile and stumble. I have to be careful but I do live alone--he says I can for now. And then on top of all my problems I have a muscle spasm, so I'm kind of nervous. Then I had this spasm in my throat which I have to see another doctor for tomorrow.

But my dad had it and they didn't know what it was in '62. He died from it, so I had my doctor check over at Colorado General about my dad's records on it, because I had my dad's body turned over for medical science. But they couldn't find any records on my father--he couldn't find any records at all of what had caused my dad's illness or anything.

And I know for a fact, I've seen my dad taken away, like that one lady said about her husband, having to be handcuffed and taken off. Well, my dad was. And I've seen him in a mental institution. I've seen my brother in a mental institution. He was in there from 16 until he died at age 32. He died in '73. He was in a nursing home and I saw him go.

And now I got real bad last year after I was told this and I got down to where I almost wanted to commit suicide myself. I got so bad I called my mother up and I told her I wanted my grandma after her. I was told my grandma had passed away. So I got so bad one day, I had a nurse--anyway, I got so bad I called my mother up and I told her I didn't want to live anymore. She called my nurse and she came with another psychiatric nurse. And she calmed me down and told me that wasn't the way to be. So, I got thinking and I talked to her all that day. And now I've got it that I want to live

my life from one day to the next. And I wrote a poem to my nurse on one day at a time as how your life lives, what makes you happy and makes you sad. She said it was so sentimental she turned it over to her boss and her boss was really enthused about it. So that's how I am now. I'm just going to live one day to the next, but I have problems with my children, and my sister, because they won't understand and everytime I try to talk to my sister about it all she says is that I want sympathy. My kids tell me I don't have it, but they don't understand. So now I have a problem--will my children inherit it or not--I don't know.

GUTHRIE: I want to say something. We've come to an end of a long day, and I want to tell you something. I was in San Francisco the other day and happened onto a television program. It was about a young man who didn't want to live and he left a note and the note is very appropriate to what you said.

His note went something like this: "Living means something to do, someone to love, and something to look forward to." I believe that we can answer all of those needs if we try really hard together. There's an awful lot to do which this Commission has testified with you today. There are a lot of people to love, and they're not necessarily our blood relatives, but people who care and want to care about each other. There is a great deal to look forward to if we succeed because it means that what we're doing today may make it possible for all our children to have tomorrows. So that little note that he left made me think of this and I wanted to say it to you and I would like to remind all of us. I've come to the conclusion--and I make my own quotations--life is a fatal disease, and being born is the first symptom. With those words I say to all of you, we can talk, those of you who would like to stay--Nancy and I are here.

Some of you I know, are very tired, but I hope you will remember what we're saying. It would be a shame to die when there's so much to do. And the price of living is to accomplish what we want to accomplish. So for every one of you, and for all your friends and neighbors and relatives, we have to reach out to the community. We have to tell them what our problem is, so that they can help us. You heard me say earlier in the day no research can be done on ghosts; it has to be real people and we have to be strong and step forward and say, yes, here I am. What can I do to help? And running and hiding wouldn't have brought us to where we are today if we had still been running and hiding. But look how many people did come forward and Nancy and I have been now in eight cities taking testimony and seeing people just like you coming before us, speaking out; we know that some of you found it very difficult and we thank you. And we know that you've helped because you came. So, let's look forward--let's meet again, and let's make a difference by living, not by dying.

Nancy, do you want to add anything?

N. WEXLER: How can I add to that?

GUTHRIE: Thank you all for coming.

Denver, Colorado

April 19, 1977

BRENDA AITKEN
DENVER, COLORADO

MARCH 31, 1977

I am a member of an H.D. family. My mother, now 48 years old, was diagnosed as having H.D. in 1972, the day after my high school graduation. Her brother, age 46, also has H.D. He has three children that now are at risk of having H.D. My mother has two daughters at risk.

My father was forced to face the fact that mother was ill in 1971. She had made several threatening phone calls to relatives. She must have been ill for several years prior to our acknowledgement of the illness, but we had refused to face the fact. Her personality had taken on a schizophrenic appearance, with extreme violent tendencies. Life must have been very threatening for her.

Father obtained help through the Tri County Mental Health Center in Adams County, where she was first diagnosed as being schizophrenic. We went to different sessions at Tri County as a family, seeking help for mother. She did not seem to improve but instead worsened. Dr. Larson had her admitted to Fort Logan Mental Health Center for about one year because we were unable to control her at home. There she was heavily sedated. Dr. Larson changed her diagnosis to H.D. in 1972 after tracing the family tree.

Mother is now in the Plaza Care Nursing Home. Father has a very serious heart condition and is unable to care for her at home. We visit her from time to time, which I have to admit isn't as often as it should be. It totally destroys me everytime I visit her to see her in the condition that she is in. She has no control over her excretory system. Her speech is drastically impaired and of late has again taken on some violent tendencies. Father used to take her home on weekends, until she was no longer able to go home. Now he doesn't visit her anymore because she has, on numerous occasions, been bodily carried from his car by the nursing staff, for wanting to go home. I don't know if mother ever really accepted this illness. As long as she was able to talk to us she denied that she was ill. Her younger sister, who has not yet shown symptoms of H.D., still refuses to believe that mother is ill and blames us for having her placed in a nursing home.

As for me, being at-risk of contracting H.D., I have learned to live from day to day not planning my future too far in advance. Unless a method is discovered soon that will: (1) determine if I am an H.D. victim, or (2) enable doctors to detect this dread disease in a fetus, I am afraid of having children. At this time we have not decided whether to have children or not. That is a decision for the future. My only escape for this problem right now if we should get pregnant without planning to is legalized abortion. This in itself will be a problem getting my husband to agree to.

I pray daily that a cure will be discovered. The fear of having H.D. is tremendous. I have found that in order to live with this fear, I must set it aside in my mind and live from day to day. Each time I visit my mother, I leave with the wish that euthanasia will soon be legalized. Her prognosis for life will be eventually fatal. Euthanasia would remove the dreadfulness of the disease. She could instead just go to sleep very peacefully.

HAROLD M. ANDERSON, PH.D.
BOULDER, COLORADO

MARCH 22, 1977

Although I have worked in the area of Science Education for nearly 40 years, only during the last few years have I become aware of the variety and seriousness of the many diseases that are transmitted genetically. Too often our endeavors of teaching genetics in secondary schools have been restricted to eye color, intelligence and the traits of peas.

Teachers do not intentionally avoid the subject of birth and health defects that are genetically transmitted, but many are poorly informed on these topics. Limited material is available that has been prepared specifically for students in science, health education or family living courses.

Materials such as the sound/slide program "The New Genetics" produced by the Biological Sciences Curriculum Study are urgently needed. Guidelines and educational experiences to increase the awareness and sensitivity of teachers are also needed. Materials on specific diseases such as Huntington's are sorely needed at the level of comprehension of secondary school students.

Therefore, I urge this Commission to seek support for the preparation and dissemination of educational materials of the following types:

1. Materials developing an overall sensitivity to the problem that genetically transmitted disease and defects and the importance for prospective parents to secure current medical advice, study of family history, and testing, if available, prior to marriage.
2. The preparation of disease specific materials for high school and junior college education. These materials must be prepared by experts who stress identification and the prevention of further transmission.
3. There is a need for materials for use in preservice and inservice teacher education programs to assure teachers become aware and sensitive to this problem.
4. After these materials have been prepared, wide dissemination will be necessary.

DR. DONALD ARMSTRONG
DENVER, COLORADO

MARCH 21, 1977

As a research scientist, I wish to express my interest and concern that adequate funding be provided by the United States Government for a program aimed at the eventual extinction of Huntington's disease (H.D.) in man.

I have been working for the past three years on how certain enzymes might be involved in the etiology of H.D. Specifically, we are trying to find an enzyme assay on white blood cells or skin tissue cultures, that would enable us to offer a laboratory test for diagnosing H.D. before it is clinically evident. Preliminary results indicate that such testing is possible but studies are progressing agonizingly slow because of a lack of funds which would enable us to hire full-time research personnel. Since whatever funding presently available comes to us from private organizations, all of us who are trying to work on H.D. are equally affected. I am certain that many capable researchers are interested in H.D. but are not willing to attempt studies on a part-time basis. The various reasons for not wanting to become involved in such a situation is obvious.

Many letters will be directed to you describing what H.D. is, how it affects the patient, his family, and others who are involved in diagnosis, treatment and counseling. Statistics are available to indicate the large number of people this encompasses. Therefore, I will not dwell on these well-known facts but rather by way of this letter I should like to propose a few ideas I feel would have beneficial effects on the ultimate goal of eliminating H.D.

All of us in biomedical research are keenly aware of the reduction in assistance from the Federal Government over the last several years. The rising cost of administration and the necessity for balancing the budget makes it somewhat easy to partially understand why this has taken place. Of the funds currently available however, none are specifically designated to study H.D. or related areas of neurological disease where pre-senile changes occur. Federal funds allocated for this purpose would certainly assist in an early solution to this problem.

In an effort to consolidate and conserve expenditures, funds could be provided to study other related neurological diseases in conjunction with H.D. where there is an appropriate overlap. Other ways to reduce overall expenditures would be to institute a series of regional workshops where scientists can convene to discuss and share their ideas. Hopefully, several investigators with similar interests would band together and work on certain aspects of a general project in a real collaborative effort. I have done this very thing and have found it to be very efficient in terms of time and information gained.

I would also recommend the establishment of clinical research centers where patients could be admitted for predetermined periods of time and be studied by all interested professional personnel while at the center. I regularly use such a clinical center here at our medical center, and can speak with experience for its effectiveness.

In summary, I see a critical need for financial support to the medical research community for 1) studies into the cause(s) of H.D., 2) identification by laboratory methods of patients at risk, 3) genetic counseling, and 4) ultimate therapy. What is learned from this form of pre-senile dementia will surely be valuable in understanding the other forms of early age-related changes of the central nervous system. I strongly urge the Senate to approve legislation to support medical research, specifically involving H.D. but also to include appropriately related studies.

Denver, Colorado

April 19, 1977

MARTHA BARNETT
WESTMINSTER, COLORADO

MARCH 23, 1977

My mother, sister and brother (from my immediate family) have lost their lives to Huntington's. Another sister has all the evident signs of Huntington's and five more of us sisters are "at risk." My mother's brother committed suicide, one of the effects of Huntington's disease, and one of his sons passed away in a nursing home in which he had been placed--although cancer too, his life before Huntington's had run its full course and reached its ultimate conclusion.

We first learned of Huntington's chorea in 1956 when it was diagnosed in my mother, who was then 55 years old. It was also diagnosed in my brother, 36 years old at the same time. This was the first time we had ever heard of the disease. We were forced to put my mother in a nursing home where she passed away at the age of 61--a completely helpless person, unable to communicate, feed herself, or even in most cases, unable to swallow her food.

My sister-in-law had to put my brother in a nursing home, so that she could support herself and the child still at home. They had three sons who are now "at risk." My oldest sister ended up in a nursing home also, and both of them have since died as a result of this horrible disease. My sister had only an adopted son, who does not have to worry about Huntington's disease--except what it did to his mother. His two children are not at risk.

Unfortunately, most of us already had our families before we ever heard of Huntington's disease and its terrible consequences. Now, this insidious killer has a possibility of 45 victims in our family alone, i.e. my siblings and their sons and daughters and grandchildren.

Some of our children have decided against having children while others have decided to take the chance that their parent will not have the dreaded disease.

It is a tragic thing to watch vital, strong people waste away and become vegetables before your very eyes and face the uncertainty of when and/or if it will happen to you. Or, what sister or cousin will be stricken next.

My plea to you is for funding to research Huntington's

disease to find a cure so that my four children, two grandchildren, sisters, their children and grandchildren, along with all the other Huntington's disease families may find some peace of mind in this world.

I also would like to recommend a health care center for H.D. patients in our area and specialized training for doctors and nurses in the field of Huntington's.

Denver, Colorado

April 19, 1977

MARY BILLINGSLEA
LITTLETON, COLORADO

MARCH 11, 1977

I have seen the effects of H.D. firsthand, because my brother-in-law is in the V.A. hospital in Waco, Texas, with it.

The doctors there can't even treat his symptoms, much less effect a cure. They simply don't know enough about H.D.

Ed developed H.D. some 13-14 years ago. His mother died in a state hospital. Ed became unable to work about four years ago. He has lived with the fear of H.D. since he first realized he had it because he saw his own mother's deterioration. Now his two children face the same uncertainty, not knowing whether they carry the gene or not.

So little is known about H.D. It will no doubt take massive research to learn how to treat it, cure it, and, God willing, prevent it.

Right now all that an H.D. victim can look forward to is years of frustration with a brain that seems to have a short circuit to the muscles and to reasoning--and eventually being put in a psychiatric ward to die. There are so many gruesome diseases in this world, but H.D. seems to me the most fearful and the most frustrating for victim and family alike.

My sister can't take care of Ed at home, and the hospital he is in is probably as good as any. Even so, about the best they can do is give him custodial care while we watch him die by the inch knowing that he really can't understand why he can't live at home. I guess that's the biggest heartbreak of H.D. If Ed were totally out of touch with the world, it wouldn't make much difference where he is. Or if he could reason enough to know that it's impossible to keep him at home, maybe he could accept where he is. As it is, he knows where he is and hates it and can't understand why he has to be there. He isn't able to reason, so he thinks he's been abandoned.

I guess it's too late for Ed, but I pray to God that medical science will find some way to treat H.D. before his kids are much older. So little research has been done on H.D. I know that you can't always equate money and cures, but at least the people with H.D. and those who love them should be given help in research and treatment.

Denver, Colorado

April 19, 1977

CINDY BOWMAN
EASTLAKE, COLORADO

MARCH 24, 1977

My name is Cindy Bowman. I am twenty-one years old. Huntington's disease has affected my life.

My Mother and Aunt are victims of HD. I'll start back as far as I can remember. We used to go back to Denton, Nebraska to see my Grandmother and family. Grandma would just sit strapped in a chair watching us and trying to talk with us, which had little results. When Grandma died, they said she died of pneumonia. Nothing was even said of her "strange mental condition." Why? Because, especially then and even now little is known about HD. When I got to the age of twelve or thirteen Mom started doing funny things. Everyone thought she was going crazy. My folks got divorced and things got worse. I suppose better for some, but worse for the rest. Mom started to hop up and down (so to speak) when she'd walk, causing my friends to laugh. She'd sit in a room for hours talking away to herself. When I'd go to visit my father my mother would get frantic. On one occasion she attacked me causing me to have to hit her to get her away from me. I'm sure at this time she was wondering what the hell was going on with herself as well as the rest of us. At the age of fifteen my father got custody of me because my mother was considered incompetent and I was unable to cope with the situation any longer. (If it hadn't have been for my sister disciplining me those three previous years, beings my mother couldn't, it's hard to say where I'd be today.)

We then took my mother to Colorado General Hospital. (She had been to previous doctors, but no one knew what was wrong with her.) They ran tests and diagnosed Huntington's chorea. My mother would cry, saying it would be her fault if we got the disease. Mom stayed home for another year and then after the death of her oldest son and our brother, and the disease grasping on to her we had to put her in a nursing home. She was in the same one for four years when she was given an overdose. It was a murder attempt, ok maybe the sole giver thought it was a mercy attempt, because then as now (about a year later) she weighs about 55 pounds, can't walk, feed herself, or talk. She just lies in her bed jerking quite often (less now than before). I feel she knows when we are there, but it's so hard. What do you say to someone you love so much when they can't communicate with you?

There is a definite need for more research on HD. I don't think I will ever marry or have children if something is not done, because I couldn't put any human being through what we've been through. If I myself do get it and there is no cure I don't think I'll wait around somewhere not knowing what or where reality really is. I'm really interested to see how the hearing in Denver turns out. If there is anything I can do let me know.

Denver, Colorado

April 19, 1977

ELLEN BARNES BREESE
PHOENIX, ARIZONA

DECEMBER 8, 1976

All my life I have been associated with H.D. and one thing that stands out more than any other is that no two people are affected the same or have the same reaction to the disease. My grandfather contracted it about the time I was born, and when I was 13 he died quietly in his bed. He was the only one in our family that died a natural death. At that time it was thought that he had St. Vitus Dance; we hadn't heard of H.D., of course. The last year of his life he was getting quite helpless and the last six months he had to be fed and cared for completely. He had five children and the two oldest, one of whom was my mother, had H.D.

The older of the two, my aunt, lived in a small mining town and her first symptoms were a mania for cleaning. Then she started saving food, even spoiled food, for when her grandchildren would come. She was always a very religious woman but as her mind changed, she became more violent and cursing those who opposed her. Her children tried getting her to a doctor but she refused any help at all. Finally, they took her by force, which was a very unpleasant task, and the doctors recommended the state hospital, where she died six months later. She had two children and neither of them have H.D.

My mother, also a very religious woman, started getting it in her early fifties, but she was not a person to let it get her down. She accepted the fact that she had it and started doctoring from the first. She continued to go to church, and every week she would ride the bus across town to my beauty shop to get her hair fixed. Then we would spend the afternoon visiting her friends and relatives in the area, before taking the bus back home. She had quite a bit of movement, so naturally we worried about her crossing the streets, but figured getting out was worth taking the chance. She was in a bad automobile accident and spent two months in the hospital before she died; the last month of which she blanked everything out of her mind. We think that was her way of getting away from the pain. She was the mother of eight children, three of whom contracted the disease.

Her first child, my oldest brother, was the first to show signs of H.D. He was a farmer so was able to carry on with his work, longer than if he had been on a job, and he had many falls

in the ditches. He also went to a doctor for treatment, and when he started showing violent temper spells, the doctor persuaded him to go to the state hospital on a six-month voluntary program to see if they could help him. They not only couldn't help, but because he disliked being out there so much, I'm afraid it did him more harm than good. So when he was released it was hard for him to get along at home. They were at the point of recommitting him when he died of a heart attack. He had five children; one he lost as a teenager and his oldest boy is showing definite signs of H.D.

My youngest brother was next to show signs of having it. He was a barber and it became increasingly hard for him to hold still. We tried to persuade him to go to the doctor but he refused to admit there might be something wrong. As he got worse, he finally was persuaded to go to a neurosurgeon in the St. Joe Hospital. He diagnosed it as H.D. and sent him to a doctor that specialized in H.D. The medicine kept his movements down, but seemed to depress him. He sold his shop and started drawing medical social security, but sitting around home, with the noise and confusion of children most of the time, he seemed to withdraw more and more. At times he was as alert as anyone, and then there were the spells of depression. It was during one of the spells one Sunday morning he got up and dressed to go to church and then went outside. His wife missed him and started looking for him, but he was nowhere in the area. So they got the car and drove around looking for him, and not finding him, went back home to see if he had returned there. It wasn't long before the hospital called and said that Dudley was there and had an accident, and that his wife was to get my other brother to go with her.

When they arrived, they found out he had jumped from the eighth story window. We believe that he went there with the intention of getting Dr. White to help him and when he got there, it being Sunday everything was closed and dark, and he must have panicked or else he couldn't find his way out and forgot he was on the eighth floor--we'll never know. Anyway, they called the Foundation and they came right out and removed his brain for research purposes. He was only 49 when he died and left three children of his own and three adopted children.

I have a sister who is still living; she's 58 and has H.D. She was always sure that she would be the one to get it, so because of it she never married until in her middle thirties, and then never intended to have children; but she has one child about 23 now. The medicine they gave her really overdrugged her; she was like a zombie and also it affected her throat to where she couldn't swallow her food. So they took her to the hospital

and they decided she was allergic to the medicine. So now all she takes is valium and she has quite a bit of movement, but her mind is clear. She is a recluse though and will not go anywhere people will see her and only lets the family come visit.

After Dudley's death, we had two doctors from Denver, Colorado come here to get the geneology of our family and take blood, urine, spit and other small tests to determine our line of heredity. They were most happy to get samples from my sister who still has it.

So you see, I have seen nearly four generations of H.D. and I personally believe that we should live a normal life and not live in fear of it, for through the tests and work they are doing now, maybe the fourth generation will be able to receive help. One of the happiest days of my life was when I learned that it does not skip a generation and that my three children, twelve grandchildren and one great-grandchild so far will never have to worry about getting it.

Denver, Colorado

April 19, 1977

VIRGINIA BRINK
DENVER, COLORADO

MARCH 21, 1977

I just found out about this Commission last week. The volunteer I talked to recommended that I write to you to let you know how I felt about the spending of whatever funds may be needed by some families to relieve the financial burden caused by this disease, I would hope that most of the funds could be used for research. I think there are many welfare and social programs available now to help people in financial need although there may be some obstacles.

My mother died of "HD" in 1963 and right now my 30 year old brother has it. I have a younger sister and brother who are "at risk." I am 36 years old and am also "at risk." Our family was literally torn apart by this disease, and I hope research will someday find a cure.

Denver, Colorado

April 19, 1977

PAULA BRYAN
PHOENIX, ARIZONA

APRIL 15, 1977

I am a single female, 39 years old. My mother died two years ago of H.D. after a 15-year illness, 13 of which were spent in a nursing home.

I was diagnosed as having H.D. five years ago and had to stop working 1-1/2 years later. Of course, I've had many financial problems, my only income being Disability Social Security of \$223 per month. My rent is \$90 per month, and naturally my apartment is not nice or comfortable. This leaves me \$133 per month for food, clothing, dental work, drugs, cosmetics, etc. I've always loved coffee, but was forced to give up my favorite beverage a year ago because I can't afford it.

Even more of a problem for me than finances, however, is BOREDOM. Since the chorea movement has become quite pronounced, no one will hire me and my only recreation is going to the library and CCHD meetings.

We've all read that "suicide" is a "symptom" of H.D. I've tried it. Let me tell you why I tried to take my life:

I had met a gentleman who I really liked, and we flew to California where we spent a wonderful 30 days together. It was my first flight and we did a lot of sightseeing--and I was happier than I had been for years.

Then it was over, and he had to leave. Alone in the motel room, I realized that my disease is progressing rapidly and that I undoubtedly would never be able to have another such vacation. Quite simply, I decided to end it that way before I got worse. I did not want to spend years in a nursing home in the condition my mother was in, so I took 52 sleeping pills.

I wasn't successful and therefore spent two months in a mental hospital before being allowed to return to Phoenix. So you see, I don't believe suicide is a physical symptom of H.D.

I hope the Commission will recommend efforts to create some way in which we H.D. victims can do creative and useful work for as long as possible. Also, better education of the public might help in the area of our being shunned and abandoned.

CYNTHIA CARPENTER
LEADVILLE, COLORADO

APRIL 3, 1977

Every single day the fear of H.D. flows through my mind for I, too, am at risk. I have two daughters, ages 13 and 15, who also are at risk and some day soon, I am going to have to tell them this. I am 35 and at this time have no signs of H.D. but every muscle twitch brings it to mind. My father died at age 70 of H.D. after suffering for at least 14 years. There were four children in his family, two died of H.D. and two lived normal lives. Our family history is not typical of H.D. in that my paternal grandparents lived to be quite old with no trace of H.D. However, the doctors feel that my grandfather must have had a mild form. He died at 68 with prostate cancer. His mother died very young with the cause unknown. She had just one child. His father remarried and had several children, none of whom were diseased. That is the only history of H.D. in our family.

My first experience with H.D. was as a teenager when my aunt came to live with us. She would get violently mad at me for no apparent reason causing great mental strain on the whole family. As she got worse she was able to be placed in a research center for H.D. at the state hospital in Rochester, Minnesota under the care of Dr. Pierson. She choked to death on a piece of meat, ending her struggle.

My mother was able to return to teaching to support our family when Dad was unable to work. Dad stayed at home for many years which, of course, caused many problems for all. I had married and moved away from home. He would be so frustrated about being "worthless" and being a "burden." I think his mind was very accurate for many years. When he could no longer take care of his bodily functions, Mother placed him in a nursing home. His care was not adequate because no one knew the problems of an H.D. patient. Most of my mother's income went for his care and in later years Medicare or Medicaid helped some. He wasted away--my strong father who was so full of fun and life--and no one could relieve his suffering.

My sister is 31 and has no signs of H.D. She and her husband have chosen not to have any children. I found out the definite diagnosis of my father's illness when I was pregnant with our second child--too late for me to make that decision. Now I must tell my daughters that they may have to make the

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same choice someday. This haunts me as does the fear of having it myself and passing it on to them. I will not be a burden to my husband and daughters, medically or financially. They will not see me waste away like my father did.

I am asking you members of the Commission to request from our Government the money necessary for a concentrated research program on H.D. for a prevention and CURE. We have to stop this disease. A discovery of medicines to treat H.D. patients would be a tremendous accomplishment. We need nursing care facilities for H.D. patients with qualified staff to help care for them. Financial assistance for their care should be available. I feel that most money should go into research.

Our Government takes care of people who do not want to work through welfare. Won't they take care of those who have led honest lives, always paying taxes and being good citizens but having the horrible misfortune of H.D.? Won't you please help us?

Denver, Colorado

April 19, 1977

ANN M. CHESTER
PHOENIX, ARIZONA

JANUARY 14, 1977

As a Probation Officer having had a client tragically sinking into Huntington's Disease, I urge you to do all you can to persuade Congress to allocate the funding to forward the research necessary to wipe out this killer of people in the prime of life.

I am familiar with the work of NIH, having been a patient there about twelve years ago. My life was saved by a study experimenting with a new medicine. Today, as a result of this good work, I am a productive member of society, am seeing my children through, and helping others to reconstruct their lives. If anyone can help, NIH can.

Please do all you can.

Denver, Colorado

April 19, 1977

MARIE L. CIVIC
MESA, ARIZONA

OCTOBER 26, 1976

I am writing in regard to Huntington's disease. My husband was told he had it. We never knew or heard of this disease. He had been sick almost thirteen years. Only our God knows what they go through; life wasn't easy. Oh how he tried to help me. I took care of him at home because I couldn't afford to put him in a nursing home. The doctor I had told me I couldn't keep him in a hospital as there was no room; but I saw a lot of empty beds. I never saw anyone with this disease so I tried to do the best I could; if only he would get well. But he grew weaker and less mobile until finally he was bedridden. I washed sometimes six to eight loads of clothes. Sometimes I thought there was no end. Besides wash, change him, dress him, I learned to shave him and feed him. Once he saw all the work I had so he got out of bed and fell. He said he wanted to help me. I told him not to bother as he could have hurt himself and it wouldn't be worth it, and not to worry if I didn't do it one day--I'll do it the next. But I thanked him and gave him a kiss. I tried to do the best I could for him, but it was getting harder and harder. But he was grateful for everything I did for him; always said thank you.

As for doctors out here, forget it. I had to learn everything for myself. At the last hospital he was in, they did very little. When I came I changed the bed to make him comfortable and had to tell them what I fed him as they didn't seem to know anything about the disease. So it would be nice if the Government could help the people that are or will be in the position I was. As for me, my darling husband died August 9, 1976. God thought he suffered enough so he took him. I miss him very much. We would have been married 42 years on September 8, 1976. So whatever you can do to pass a law to get some money from the Government to help other people, I am sure it would be appreciated. Thank you and May God Bless You and Yours.

FAMILIES OF PHYLLIS AND TOM CLEMENTS
PUEBLO, COLORADO

MARCH 21, 1977

SPECIFIC PROBLEMS AND RECOMMENDATIONS

Tremendous ambiguity surrounding Huntington's disease requires a great deal of treatment flexibility. There are so many interacting factors, both physical and psychological, that it is nearly impossible to specify at any given time what is causing patient behaviors. Medical and treatment plans are in most incidents experimental and rarely stable over time. The uncertainty felt by medical personnel is experienced 100-fold by the family.

It is critical that family understand reasons for the variable nature of treatment and are informed as to results. Changes in treatment are bound to be unsettling to both patient and family and should be handled as smoothly as possible. In addition it is important that continuity of treatment responsibility be maintained. In good faith, many physicians will refer a Huntington's patient to another physician whom they feel could be more helpful. Given the general lack of knowledge about Huntington's disease, a referral is generally not helpful and results in more confusion. The patient and family are then faced with having to develop a new set of relationships with the new treatment staff.

As effects of Huntington's become more pronounced, radical changes take place in the dynamics of the family, in particular, the marital relationship is highly stressed. It takes a very sensitive staff to recognize these changes and effectively work with them. Although hospitalization is inevitable, eventually, upon diagnosis, it is impossible for the spouse to fully prepare for it. Strong ambivalent feelings are bound to be expressed and should be recognized as such by treatment staff. Difficulty in making clear cut decisions should not be interpreted as lack of cooperation by patient's family.

Following are more detailed responses to your questionnaire. They are purposely at times more detailed and verbose than you may deem necessary, in hopes of better illustrating the "nitty gritty" particulars that formulated the above observations.

I. What are the specific problems and recommendations associated with being "At Risk" for H.D.?

1. Additional stress and the lack of a stable environment during formative years, particularly teen transition. Independence and responsibility are forced on them earlier than most, while creating dependency at the same time, because one parent is having to secretly do their best to supply the emotional support that the other is knocking down faster than it can be rebuilt.

Recommendation: Provide counseling service to help both parent and children involved understand what is going on, and formulate progressive steps to maturity.

2. Need to provide the parent that is coping with a source to compare notes with. Phylis's "therapist" was her hairdresser, where she found other mothers of teens to talk to, discovering many times that her kids were pulling the same stunts as everyone else's. The loss of ones spouse as a sounding and decision making board, is similar to one experienced by a single parent, only twice as frustrating, because the other partner is there, not able to be relied on for rational decisions and in many cases, unwittingly thwarting every effort.

Recommendation: A professional service may not be necessary for this aspect, but the availability of a personal contact volunteer that the parent can talk to by phone or in person. Somewhat like the Single Parent Organization, only composed of parents of non-risk children. Association with Huntington's groups will help parent form guidelines as to what other "At Risk" children are going through.

3. Premarital Stress and Decision Making. Knowing that they face Huntington's, there are additional factors to be considered: the willingness of mate to live with the constant threat of H.D. and undertake care if partner does become victim; make major decision on children

(whether to have own and gamble on the future, plan to adopt in an ever tightening availability market, or not to have children at all); how to prepare financially for the future "in case."

Recommendation: Adequate information and counseling on which to base these decisions. Couples should know what the options are in the area they plan to live. Example: In Pueblo, Colorado, both private and public adoption agencies place both adoptive At-Risk infants and infants with an At-Risk parent on a flexible, individual basis and evaluation according to the strengths of the couples involved, especially the partner not At Risk.

4. Legislation Needed So that Huntington's families do not have to face the additional stresses of splitting up the family by divorce in order to receive medical and financial assistance. Also to provide access to sealed adoption records to confirm diagnosis in suspected Huntington's victims who are adopted.
5. The only realistic hope that exists for the At-Risk group at the present time is through discovery of the gene involved and measures taken to replace or correct its destructive force. Volunteer victims and At-Risk people are essential for the success of this program. Yet, our children are so embittered by their research experiences that they refuse to cooperate in further testing programs.

Recommendation: Keep volunteers informed of all aspects of testing involving them. What for; how test will be conducted; why this particular test; will test prove theory or be basis for new area of exploration; possible side effects.

Follow-Up Check to see what, if any, after effects volunteer encountered and advise them of results of their tests. Help volunteer feel that they are valuable assets in research program, not guinea pigs to fill up test tubes.

Interviews. Try to keep all interviews on a professional basis. Living with Huntington's is a chain of unending crises and emotions riding close to the surface. One finally in order to cope finds a spot or way within themselves to "put" their fears and confusion and go about their daily living. Every crisis, interview, or research project opens up that spot and the volunteer has to work their way back to their comfort zone again. Our children were approached by a member of a research team to give her child interviews on how they felt for a school assignment.

Notation: As of this date, two of our four children are faced with hard decisions because of the Huntington's factor in their lives. One is in the process of divorce and the other contemplating marriage. Of the other two, one is successfully married, and the other unmarried. Two of our children also have one child each.

II. What are the problems and needs associated with the period after onset of symptoms prior to diagnosis?

1. Emotional support needed for person involved; protection from police to avoid arrest for public drunkenness; job jeopardy; peer group pressures; family faced with coping with accusations (our children were involved in many fights defending and protecting their Dad's integrity).

Recommendation: Focus on public awareness.
Immediate distribution of information and education of all public service related personnel is essential.
Avenue open to victim for purging negative comments or records of firing for alcoholism, or drunkenness etc, from their work record after diagnosis.
Financial assistance for those involved in job loss or inability to work prior to, or during diagnostic process.

III. What are the problems and needs associated with the period during and after diagnosis?

1. In our case, Tom suffered a tremendous loss of ego and identification as a man. The adjustment from the loss of his job and outside involvements, to being a dependent non-contributing member of the family was shattering. His regular Marine pension waived because he was eligible medically for Veterans Association and Social Security benefits. It took over 18 months to process these. Because I worked, we were budgeted for two incomes. When one was suddenly cut off, Tom felt responsible, and that he had to find a job to help make ends meet. He finally found one job that was very unsatisfactory and de-meaning and involved considerable transportation problems. It lasted less than six months.

Recommendation: A job pool list of employers or situations available to handicapped individuals that offer some type of monetary remuneration, to make the transition period more gradual and the victim feel less dependent, without jeopardizing benefits. Any job will be extremely temporary.

Counseling to help the victim regain their perspective as a valuable human being.

Immediate steps taken so that any pension granting facility recognizes Huntington's chorea and its effects on a person's ability to work, so that processing can begin upon verification of the diagnosis. The family is less jeopardized financially, and the victim feels less dependent when something he is responsible for is contributing to the support of the family. The lack of information about Huntington's is what held our benefits up for 18 months.

2. Change of legal status. A need for competent legal and insurance advise, to set up wills, trusts, power of attorney, review coverage in light of diagnosis etc,

while victim is still able to make wishes known and objective decisions. We also made our burial plans, so Tom's wishes could be followed when the time arrived.

Recommendation: Pick insurance agent and lawyer familiar with, or close enough to, family to understand victims' instability. As disease progresses the victim may frequently change his mind and demand that changes be made. The professional involved will have to be creative enough to alleviate the victim's concern, and maintain status chosen by both partners when victim was still able to make objective decisions.

3. Need for family awareness.

Recommendation: Counseling and information for all family members who are closely enough tied to the affected family to offer definite emotional and social support and cope with the victim's moods and tempers without being subjective. Additional quarreling and bickering pressures are not what victim, spouse, or family needs.

4. Loss of friends, both of his personally and ours as a couple, when we needed them most. They couldn't handle our new situation, leaving us virtually alone. Both of our families lived over a thousand miles away. Frequent phone calls and "fly-ins" by various family members served as our emotional support.

Recommendation: Leaflets available to pass out to friends to explain in abbreviated form the disease and the social needs of the victim and his/her family would be helpful. Anyone who is willing to cope with the situation can be given more detailed information and included in counseling sessions where appropriate.

5. Danger from police if anywhere alone because of staggering, swaying movements and slurred speech.

Huntington's victims are easily mistaken as drunks and arrested.

Recommendation: Medical alert bracelets or other form of identification for victim to carry on their person to avoid public embarrassment and detainment. Note: In Pueblo, alcoholism is recognized as an illness, and suspected drunks are taken to their homes or a medical facility for treatment.

Identification is also needed for the safe return of victims, who also have a tendency to wander off and get lost.

6. Campaigns to educate the public about Huntington's and to help others understand the need for Huntington's families to retain their social contacts and carry on in a normal manner for as long as possible. The medical, financial, psychological and social blows hitting all at the same time are almost more than one can take. Most people who withdraw from people afflicted with "strange" illnesses do so out of their own lack of knowledge and fear of doing the "wrong thing" when in the presence of the victim or his/her family.

IV. What are the problems and needs associated with the period during which the patient can remain at home but needs increasing support?

1. Household help, or facility for outpatient care while the spouse is at work.

Recommendation: Possible day care at already existing facilities like nursing homes, where victim could have some social contact as well as supervision.

2. Support and regular visits by family and friends. The outside coming to the affected family.
3. Visiting nurse service to free spouse or administer or monitor special medication and needs. While this

service was available in Pueblo, we couldn't use it. Tom would not let Phyllis leave him day or night, even for a few minutes. He would rarely consent to even stay with another family member.

4. As disease worsens, victim may feel obsessed with the need to be constantly going somewhere.

Recommendation: Know places to go where victim and spouse can feel free to utilize and maintain as normal as possible life style.
Consider architectural barriers, distance to walk, attitudes of store, restaurant personnel, social contacts who don't mind frequent "drop ins," and the flammability factor for smoking victims.

- a. Handicapped parking is available in Pueblo, but Tom didn't quality because he was "ambulatory." His walk was unsteady, unreliable and not good for distance, but he could walk. Availability of information on Huntington's would have enlightened this particular decision making board.
5. Know where to turn for domestic help. Take any normal phase of home maintenance and magnify it about 10-fold. The supervision of the victim becomes so constant that there is, literally, no time to spend on numerous phone calls or trips to remedy inevitable home crises or needs. One is not emotionally or physically up to tracking down resources.
6. Recognition of the difficulty of victims and their families to unite for better efforts in research, publicity and benefits, because of the need for constant supervision and the termperment of the victim.
7. Continuing social problems. Not many are willing to accept "our brand" of socialization.
8. VERY IMPORTANT As time for residential care nears,

the need for objective, sensitive professional advice and support of involved family members is imperative to help evaluate, make, and live with decisions. This is particularly important in cases like Tom's where the mind goes before the body. The mentally ill can seem so normal at times. The final decision has to be made on an individual basis, of course, but the spouse is daily living in such an abnormal situation that they can lose sight of what normal is, and need outside feedback to help evaluate their particular situation. Both family and treatment staff should be sensitive to strong ambivalent feelings expressed during this time by the spouse, and not interpret them to be a lack of cooperation.

- V. What are the problems and needs associated with the time when residential care becomes necessary?
For us there have been many.

1. Prior information proved incorrect. The paper stating that Tom had been medically declared mentally incompetent and a phone call to the VA facility at Ft. Lyon requesting an ambulance would be all that was necessary to transport him and hold him until legal commitment could be obtained. It was our hope to make the transition as uneventful as possible for Tom's sake. Instead, Tom was transported by the police, in handcuffs, to a local hospital, where our family physician and a local psychiatrist admitted him. He remained there for a week. Part of the confusion was Tom's suicide attempt made immediate action necessary and vast misunderstanding of legal ramifications on everyone's part.
 - a. The V.A. fluctuated between sending an ambulance (too expensive), paying a private ambulance service to make the 180 mile round trip, and having him transported by the Sheriff's Dept. in handcuffs.
 - b. Would not accept Tom before legal papers were completed on anything but a volunteer basis, from which he could walk away if he demanded to.

- c. Recent changes in law concerning the rights of mental patients began a legal hassle over who and what took precedent. We had Federal, state, local laws, a time lapse factor between petition for committment and the signing of the order, and crossing three county lines from place of committment to county of residential care to contend with. It took the combined efforts of the District Attorney's office, the police, and sheriff departments, the local hospital and their social worker, our psychiatrist and family physician, Phyllis' psychologist's brother, plus another brother who has a definite aptitude for eliciting direct responses to specific questions, a private ambulance company and two judges to unravel the red tape.
 - d. Being stood up by one judge who decided to take the day off without telling anyone. That necessitated waiting another hour and a half for a judge to get free to sign the commitment paper, and delaying transportation another full day.
- 2. After commitment, Tom was shifted five times within the first 90 days to different and/or back to various wards, each time under different treatment staffs, presenting a disruptive influence on Tom, the family, and a lack of continuity for evaluative purposes.
 - 3. Tom was asked two weeks after commitment if he would like to go home on leave for two weeks over Christmas (by a doctor who hadn't read enough of his records to see the reason for commitment). That unnecessarily built up Tom's hopes, only to have them dashed; it set the family in a dither because they knew his condition hadn't improved one iota from the time of commitment. Legally a new order would be necessary for the Sheriff to transport him back if he refused to voluntarily go. This was unlike the earlier situation where he was permitted to be transported in an ambulance, and again involved crossing county lines.
 - 4. According to the explanation given to us concerning the new laws, each mental patient is to be evaluated

every 90 days, twice, then every 6 months thereafter. If found improved or deemed to be rehabilitated by a staffing, the patient may be released at any one of these points. While realizing the law was passed to protect the individual, it is unrealistic in patients with Huntington's disease whose deteriorated cells do not get better, but continue to deteriorate. The spouse is held on a 90 day string, unable to take steps to rebuild their own life or accept employment.

5. Family members were requested for interviews as part of the evaluation because Tom's institutionalized behavior is markedly different than his home behavior. Everyone sat on the alert for two weeks, waiting for a one-day notification to make the trip down. We are not on alert anymore, but we are still waiting.
6. The 90 day commitment was up February 28, 1977. The evaluation and staffing has not taken place as of this date: 3-21-77.
7. It was decided by the doctor and social worker on the ward, with family approval, that Tom could function for awhile, yet, in a nursing home situation. A nursing home closer to the family was selected. It is in Colorado Springs, in yet another county.
8. No one can be legally committed to a nursing home. The evaluation never took place, the original commitment has run out, and we do not know where we stand legally. We have been verbally assured that VA residents are told they have a choice: remain peacefully at the nursing home or be recommitted to Ft. Lyon.
9. The doctor on the last ward plus the medical director of the hospital have told us that home care for Tom is no longer feasible and not even considered as an option. Yet, his transfer to the nursing home was delayed when he decided he didn't want to go, but move in with one of his children instead. Everything stopped until the child involved could be consulted and accept or decline. He would not have been permitted to go even if the child had agreed. However, because of the new laws, a patient

has the right to make his own decisions even if he has been declared legally and mentally incompetent to do so. Somewhat contradictory and confusing.

10. Having been delayed a second time waiting for the evaluation that never happened. The staff were not able to secure the signatures of the two doctors necessary for move until minutes before actual departure. This again kept family members on a string, having assured Tom that they would be at the new place to help him get settled in.
11. The distance factor for residential care is a large one. Not only from the patient's perspective, but the families'. Frequent visits are needed to reassure patient and to assure family that the victim is being cared for in a manner acceptable to them. Education of treatment staffs at existing residential facilities as close as possible to victim's home seem more feasible for this reason than one special facility in each state. Also, financing to help existing facilities who would be willing to cooperate, provide adequate care areas for Huntington's victims.
12. Need for emotional support for spouse who undergoes much the same trauma of a widow/widower in adjusting to living alone. Knowing that he/she is permanently alone, but that their spouse is still alive, with the personhood of that loved one becoming more and more obscured by the disease.
13. The continuing personal agony of wondering if one has made the right decision, and reevaluation of alternatives in terms of where victim will be happiest and best cared for. Nursing home? Mental facility? Medical hospital or facility? Home? And the never ending frustration of not being able to communicate or interpret the victim's feelings and know for certain what they are going through.
14. Every event, move, or change in the illness precipitates another crisis. Reestablishing relationships with new treatment staffs, reevaluating patient's response to new situation, even getting their clothes gathered together in one place for them again can be

a major accomplishment and factor in patient's adjustment to new surroundings. Preparing and assuring the children. Every change makes more real to them the future they may have to face.

15. Much of the time incidents are so fresh and close to the surface for the spouse that it's hard to discuss or even to think about them without losing one's composure. The emotional impact is unbelievable. After awhile the mind can handle just so much decision making and crisis, and goes into a stall, where even meal planning becomes impossible. Concentration is out of the question and the spouse becomes frustrated and frightened.

Tom is not yet in the advanced or terminal phase of the disease, so we do not know what new problems and needs we will be faced with.

MEDICAL PROBLEM INFORMATION: In California, we did not experience any difficulty in obtaining a specialist or medical facility. In Pueblo, the only neurologist, insisted that Tom had been misdiagnosed and presented such a personality conflict with both of us that we refused him. We have relied on our family physician and psychiatrist to take care of immediate needs, and the VA for longer term medical and neurological care.

The lack of consistency and progression pattern of the disease from patient to patient has made it difficult for anyone to know exactly what to expect or what medication will be effective in any given situation. This difference also caused the fluctuation of diagnosis from time to time.

Tom was hospitalized a year before the definite diagnosis was made, and that medical team was not able to determine what Tom's problem was. A year later, a neurologist at a different hospital recognized and confirmed the disease.

The information we have received about H.D. has been sporadic. We have found it to be informative and accurate except in terms of progression and duration. This part of the disease has varied with Tom more so than in many other cases.

While experiencing much confusion with the various doctors

since Tom's hospitalization last November, we know we can rely on our local family physician, local psychiatrist, and former doctor in California for advise, specific information and assistance whenever or in whatever way we need it.

We've experienced medical confusion in both states as to who can better treat Tom. The neurologists say the psychiatrists; the psychiatrists say the neurologists. We feel they both need to, because Tom's neurological disease is affecting his mental condition.

GENETIC COUNSELING Though available to us and our children free through the VA, the counseling we received proved to be not especially useful. It seems that every decision that was made by us had to be made according to what was right for that time. The children also made their decisions according to what they felt was right for them, and in appropriate cases, right for their spouses and children.

OCCUPATIONAL THERAPY We tried it, but Tom's fine motor control was gone before diagnosis and the unwieldy action of his gross motor muscles plus his impatient personality made it too frustrating.

OTHER RECOMMENDATIONS:

1. Prepare a better questionnaire, with brief specific questions that elicit brief specific answers. Many victims and families draw a blank when faced with general questions, but can answer specific questions when asked. Because of the high saturation of emotions, lengthy, detailed answers are almost impossible to give.
2. Send these questionnaires to every known family and place ads for unknown victims. Offer the option of taped interviews, based on the questionnaire in cases where recipients are more comfortable talking than writing.

It took several composite efforts by family members to understand and respond to your questionnaire.

3. Have all known facts and questionnaire responses collected in one central location for documentation, statistical analysis, and compilation into a document

or personalized, easily read book form, divided into chapters dealing with all known aspects and phases of Huntington's. It would be for use as a general guide by victims and any medical or public related service in which they come in contact. Preferably it would be written by someone closely associated with, but not At Risk for H.D. to convey and interpret feeling.

April 19, 1977

ARDYS B. COX, P.S.W.
ARIZONA STATE HOSPITAL, PHOENIX

MARCH 22, 1977

I have worked at the Arizona State Hospital as a Psychiatric Social Worker for 12 1/2 years. In that time, I have been involved with three families of diagnosed Huntington's disease patients.

From my experience, I am impressed by several aspects of the effects of this disease:

1. Differential diagnosis and misdiagnosis.
2. The need for residential treatment (when State Hospital is the only available facility for care).
3. Problems involved in Nursing Home placements.
4. Income and eligibility for financial assistance.
5. Pressure on the working spouse and family.

My first experience was with a very successful 34-year-old executive secretary, unmarried, who showed lack of emotional control, lost her job and was admitted to the State Hospital with a diagnosis of Schizo Affective Schizophrenia, depressed. She has been hospitalized continuously since 1968. Her mother died in 1942 undoubtedly with Huntington's disease undiagnosed. Her only sister died in this hospital after years of "treatment" for Huntington's disease.

This patient has been treated on our Medical ward since 1971 at the per diem cost of \$77.67 (as of April 1, 1977). Her father pays the hospital \$250.00 per month; this represents roughly the cost of \$137,000 of which the father has paid \$24,000. No medication has been successful in giving the patient any comfort or relief of symptoms.

The emotional toll on the father is indescribable. The psychic pain of losing every member of his immediate family to disease against which he is totally impotent and his only resource for care is as impotent as he is, is beyond expression. I see the suffering in his face, in his posture, in his demeanor, and my heart bleeds. Then I view his continuing courage, his hours of volunteer work with the CCHD of Arizona, his routine

visits to see his daughter, his acceptance of his grief, and I am inspired.

My second experience with Huntington's is a very dear, gentle, middle aged man. He has no credible history of HD in his family, but has been firmly diagnosed as having the disease. I have worked with his wife for the entire term of his hospitalizations here since 1964. He was diagnosed Schizophrenia, paranoid type, on admission and treated with phenothiazines. On a few occasions he was placed in boarding homes but regressed to a point requiring rehospitalization. His wife has sold their home, bought a mobile home, and works as a switchboard operator. She pays most of his social security disability income to the hospital for his care. Her income, though barely adequate to maintain herself and her health problems makes them ineligible for any kind of financial assistance.

We have had consultations by neurologists and by Dr. Stern of Tucson. The present chemotherapy is somewhat effective in alleviating the patient's distress in the terminal stages of the disease. His wife lives in mortal fear that the hospital will request another nursing home placement, which she cannot afford and which she knows from experience will precipitate another regression. I assure her repeatedly that such a move is very unlikely. But every change of personnel here and all talk of reorganization and changes within the hospital produce extreme apprehension on her part which adds to her already too heavy burden.

Which of us, approaching retirement age, yoked with these insoluble problems, could find an inner reserve of courage and strength to face our days alone?

My third experience is with a middle aged widow, mother of two who has been hospitalized for eight years. She was diagnosed originally as Simple Schizophrenia and Degenerative Disease of the CNS. Her mother died in another state hospital as did one sister.

She was a loving and understanding mother and grandmother and a capable housewife. At the onset of her illness, her children obtained a neurological consultation but could not finance further study or treatment. The married children were concerned and involved for a number of years, but as their families required more of their time, they were not able to keep

up with both areas of interest. Letters to them are now returned to the Hospital as "Unforwardable." There have been no visits by relatives for many many months. The patient has no income, is totally at the mercy of the impersonal institution. The two children and four grandchildren have their own fears and apprehensions to live with. Where is their hope?

The needs of these victims, as I see them, are

1. Resources for diagnosis and treatment.
2. Financing for facilities other than the State Hospital when resident treatment becomes necessary.
3. Education of Nursing Home staff for caring for patients.
4. Changes in insurance coverage through Medicare, Medicaid, etc. to assist the stressed families.
5. Adequate counseling and empathic support for the "at risk" family members, as well as spouses, etc.

Thank you for permitting me this opportunity to express my concern for my suffering friends.

Denver, Colorado

April 19, 1977

GLADYS DAVIES
PHOENIX, ARIZONA

MARCH 20, 1977

On February 11, 1912, my sister Artie Keck, who was 19 at that time, married a nice young fellow, Cloyce Benedict, who was also 19. He came to our community in Kansas from his native Ohio. In due time we learned that his father and an uncle were in a mental hospital.

After five years of marriage they had a daughter, Ina Lou, who is now 60 years of age. Later they had two other children, a daughter Evelyn and a son Donald. When Ina Lou was about eight or nine years old Cloyce's behavior changed. He became cruel, especially to Ina Lou, and Artie finally divorced him. Later Cloyce was killed in a motorcycle accident.

A couple of years later Artie married Cloyce's widowed brother Guy, whose first wife had died and left Guy with three young children: two girls and a boy. After a year or so Artie and Guy had a daughter Dorothy.

When Donald grew up he seemed very talented, and had visions of becoming an architect; but when he graduated from high school, the second world war was on and he was drafted into the army. While at the front in Europe, he was carrying a box of ammunition at night and stepped in a well. He spent some time in the hospital and was given the Purple Heart. When he got out of the service, he planned to go to college and major in architecture, but he was very nervous and couldn't do the drawings like he had been able to in high school. He did start to college, but wasn't the good student he had been in high school. He married and after several different jobs, and three baby boys, he decided to go back into the service. He reenlisted and was a sergeant. His condition worsened, and while training soldiers he fell down on the drill field. Finally they sent him to the Government hospital in Bethesda, Md. They diagnosed his illness as Parkinson's disease. As he was from Topeka, they sent him to the Veterans Hospital in Topeka. He died there several years later. His oldest son is a Vietnam veteran. The second son is in a mental hospital in Topeka. The youngest son is now in the service.

Cloyce and Artie's oldest daughter, Ina Lou, is 60 with no signs of H.D. Their daughter, Evelyn, died several years

ago with what the doctors said at that time was Parkinson's disease. She had one child, a son, who is now 28 years and has no symptoms of having H.D.

Guy committed suicide by hanging himself. His three children by his first wife are all victims of H.D. The two girls have been in the hospital for many years, and son, Bob, died several years ago with the illness.

Dorothy, Artie and Guy's daughter, is in the State Hospital in Norwalk, Calif., and has been diagnosed as having H.D. She is 48. She married and has had five children. She didn't become ill until her children were grown. She has five grandchildren. All these children and grandchildren are vulnerable to H.D.

It is my earnest prayer that some treatment can be found that will prevent the spread of this dreadful affliction, and some hospitalization will be made available to those who are victims of the disease.

Denver, Colorado

April 19, 1977

MOYA G. EASTERLING
PHOENIX, ARIZONA

APRIL 14, 1977

Perhaps the most difficult step in writing this letter about a dear one who is a victim of Huntington's Disease is dealing with the question of "Is she?" or "Isn't she?" There doesn't seem to be enough travail involved in the agonizing wait for symptoms of this genetic time bomb to appear in the at-risk human being. After that follow stages of denial, differing diagnoses by doctors, widely varying medication patterns, and (in the case of my former daughter-in-law) belief in miraculous remission and cure through her becoming a "born again" Christian.

Meanwhile, my husband and I live on in doubt, concern, and unease about the future of our precious grandson, who lives with his mother hundreds of miles away from us. Is he an at-risk person or not? Under what conditions is he living if his mother is actually worsening in her HD progression? Usually a very loving and dedicated mother, is LuAnne still suffering the periods of deep depression which we saw occurring when she lived here in Phoenix for two years? Or does she still evidence irrational anger at times with which our understanding, though puzzled, grandson must cope at his tender age of six? We're not even nearby to observe for ourselves what is happening.

These are some of the many unanswered questions that families of HD patients and the patients themselves must face, particularly in the early stages of the disease. We have done extensive reading and consulted with research experts in the field. There do not seem to be documented cases of spontaneous remission. Yet, LuAnne believes that has happened to her. What can we believe?

Let me set forth some of the details of this case for your consideration. Both LuAnne's mother and her maternal grandfather died by suicide because of HD-triggered depression. They were in their mid- and late thirties, respectively, when their tragic deaths occurred. My son did not know of this family history when he chose LuAnne as his mate. Their union was short-lived, but a male child of unusual beauty and intelligence resulted.

Doctors in Utah diagnosed LuAnne's HD while she was 21 years old and pregnant with Jason. When she moved to Arizona two and one-half years later, she brought these corroborating medical records with her and contacted a leading neurologist locally who is identified closely with his interest in the field of HD. He never wavered from concurrence with the diagnosis.

In the course of her Phoenix residency, LuAnne encountered great difficulty in keeping jobs. She was trained in the assembly of electronic products which involves soldering very small components based on following a schematic design. Yet, as her local physician increased LuAnne's medication and combined "uppers" and "downers" to deal with her mood needs, she found it increasingly difficult to perform her work. Frequent absences because of illness, followed by numerous times when she had to take time off for doctors' appointments, caused her to lose five jobs within one year. The fourth position lasted less than two weeks because the company's personnel department had initially overlooked LuAnne's honest admission of having HD. She was subsequently ruled unacceptable for group insurance coverage and thus unemployable by company personnel rules.

Harried by extreme financial duress, exacerbated by periods of unemployment and unbelievably high medical costs, LuAnne finally had to seek emergency care for bleeding ulcers at a local hospital during the middle of one night in May, 1974. Her own doctor couldn't be reached, but when the admitting resident intern discovered she was an HD patient, he eagerly summoned three colleagues. For over two hours, they interrogated LuAnne and had her perform balance, coordination, and perceptual tasks for their enlightenment while doing nothing for her presenting ulcer distress.

Please understand that I recognize the infrequency of opportunities for young doctors to see an HD-diagnosed patient and their natural desire to broaden their experiential base. However, the anguish and even personal embarrassment this incident represented for LuAnne were traumatic, to say the least. It is another facet of the problem of not having readily available and nominally priced treatment centers where the uniqueness of the disease would not get in the way of helping the patient.

A day later LuAnne was admitted to that same hospital for treatment of severe stomach ulcers. Three major developments followed: (1) she learned that her insurance did not cover the expense because she hadn't been on her fifth job over one month, (2) her doctor treating the HD, who was not the internist working on the ulcer problem, started prescribing heavier and heavier medication until LuAnne became a walking zombie, and (3) she was fired from her job when she returned to work because it was a federal government contract with a mandatory completion date, so "no sick personnel could be tolerated."

By this time my husband and I had laid out over \$3,000.00 in financial assistance to LuAnne in one year. We were not in position to do so. Therefore, when we started a retail hardware business at this point in time, we employed LuAnne as a sales clerk in order to protect her in a work environment that would absorb some of her difficulties through extra effort on the parts of family members working there, too. Thus, she was paid a higher wage than we could easily afford, granted leave time whenever it was required with no income loss to her, provided at no cost to her group insurance coverage that protected both LuAnne and Jason after 90 days--even for HD, and in general made many allowances for her increasingly over-drugged condition, which resulted from her neurologist's handling of her case.

This situation lasted nearly a year, during which time marked personality changes occurred. LuAnne caused a great deal of malicious mischief among family relationships. She phantasized about the intentions of salesmen who called on our store, or about adjoining male merchants. When I phoned her doctor for an appointment to discuss these developments, he refused to answer my call and told LuAnne that she need not worry about his talking to her former mother-in-law!

In May of 1975, LuAnne quit in anger one day when being shown how to price some merchandise. She returned to Utah to live. There her doctor reduced her medication to less than one-eighth what she was getting here, and within a short time she seemed much more coherent and cooperative than she had been in months. While she still has difficulty keeping a job, she is very involved in her newly found religious fervor and appears to be much more at peace.

How long will this phase last? She has no one but us to turn to when things get worse. Will she let us know before Jason's life is too adversely affected? Does she have the blessing of a miraculous cure? If such a thing could happen, the question still persists: Does Jason have the genetic HD time bomb waiting to emerge in him?

Is HD easy to live with? Obviously, it's a nightmare. Perhaps this sharing of our family's experiences to date will add to your growing mountain of evidence that begs for a concentrated and adequately funded national and international effort to find solutions to this scourge against mankind.

Denver, Colorado

April 19, 1977

J. M. EULBERG
LAKE HAVASU CITY, ARIZONA

JANUARY 24, 1977

I am writing this letter to tell you a few things about my encounter with this Huntington's disease. I do not want to bore you with details so I will make it short and to the point.

I was married in 1959. After a few years my wife's mother took sick, thought she was crazy. But soon found out she had H.D.; died in county hospital at about 50 or 52 years old.

My wife is now going through all this; I know she has it because of symptoms of moving in a jerking motion. She will not admit to this so she will not go to the doctors. I have three daughters also and I want to help and do things to overcome this so they will not get it.

I am getting divorced now and this is not because of her having H.D. I am going to try to help her, but our marriage never was anything to rave about and my daughters will agree. If there is anything you can do to help us help ourselves, I as a U.S. citizen am asking. I am in contact with the Arizona Chapter.

Denver, Colorado

April 19, 1977

MRS. G. F. FRANCO
PHOENIX, ARIZONA

JANUARY 10, 1976

Last Christmas, our Arizona chapter of CCHD requested me to visit Arline Richards in a local nursing home. She does not write, nor speak, but is able to indicate her choice of meals by nodding when shown a card.

She has huge, brown expressive eyes and enjoys your reading the Bible and just talking to her. If her dentures were available, she wasn't using them. Her only companion is very loud and can shout when she is displeased with anything.

I mentioned taking her to Dr. Stern's Clinic at Tucson, but of course she cannot say whether she is agreeable to that next step toward medication and future tests.

Tucson is 200 miles round trip and you stay overnight.

It would be helpful if all hospitals and nursing homes knew of therapy to keep the hands working instead of just lying idle.

This is the best time for Congress to support this small minority of Huntington's patients. Eliminate the disease before it quadruples by inheritance; the choice of parents was not ours. Congress could be the best ally to rid our fears of this unnatural death. Forward funds to research in the battle for the present generation.

Denver, Colorado

April 19, 1977

MRS. HELEN HANSON
PRESCOTT, ARIZONA

MAY 30, 1977

I sincerely feel that the Federal Government should assist in the fight with Huntington's disease. My husband has been ill with it for 15 years. The last 10 years he has been unemployed, and unable to do anything. It is hard on his family to see him and understand. As my daughter wrote to her sister, this very much describes her father--

1. He's nervous and has to have the TV and radio on as loudly as possible and gets into fights when we want it off.
2. His walking--he's off balance and rocks and falls over.
3. His eating and appearance--need improving.
4. He has to go with you everywhere, which is ridiculous. And a big complaint because after Mom leaves and comes home and he's so angry and wants to know where and why he couldn't go with her.
5. He paces and paces and drives everyone batty.

This is only part of his description.

When the day comes that I can no longer handle him at home, I will have to file for divorce as I can not get financial assistance until I am completely broke. And at \$600 to \$700 a month for nursing homes--it won't take long.

We have 4 children who have a 50-50 chance of inheriting the disease. I feel help is necessary for the study and prevention of this terrible disease, as well as help to all families already stricken with it.

Denver, Colorado

April 19, 1977

WERNER G. HEIM, Ph.D.
COLORADO SPRINGS, COLORADO

MARCH 7, 1977

In connection with the hearings scheduled for April 19, 1977, in Denver, Colorado I should like to bring the following to your attention.

As a student and teacher of genetics, I have given considerable thought to the problems presented by Huntington's disease. My main conclusion may be one with which persons afflicted with the disease and their families may find it somewhat difficult to concur fully but it is this: that the main thrust of research with respect to Huntington's disease should be sharply focused on a means of determining, before reproductive age is reached, whether or not a member of a family in which this disease has occurred has the allele for the disease.

There are two reasons for reaching the above conclusion: First, if such detection is available, members of families with Huntington's disease can make their life plans with much greater certainty. Such matters as choice of a profession, marriage and reproduction, property trust arrangements, etc. all can be made better if definitive information as to the presence or absence of the allele were at hand. True, for about one-half the persons in such families the information would be sad and its psychological consequences, especially without the support of well-trained professional and lay persons, sometimes quite serious. On the other hand, however, about one-half of the family members could plan their lives in the certain knowledge that they will not develop the disease.

The second reason for my conclusion is that the development of an early test for the absence or presence of the Huntington's disease allele would permit the elimination of that allele in a time perhaps as short as one generation. If ever one who possessed this allele knew the fact and made arrangements not to reproduce the disease would, for practical purposes, be wiped out. Even if only a fraction of those who have the allele would inhibit reproduction, the frequency of the disease would be proportionally reduced. Development of an appropriate test would act, in conjunction with inhibited reproduction, very much as a highly effective vaccine might for an infectious disease.

Finally, there may be an additional benefit obtainable from successful research into the development of a test for the presence of the allele. It is entirely possible that such a test might be applicable to the fetus in utero. In that case not only would the virtual elimination of the disease be easier but also persons having the allele could safely reproduce and be assured of having only normal children if they terminated any pregnancy with a fetus possessing the allele.

I hope that the above thoughts, though they lack both originality and novelty, might help direct the work of the Commission toward speedy elimination of this most unfortunate disease.

Denver, Colorado

April 19, 1977

SUSAN HEITLER, Ph.D.
DENVER, COLORADO

MARCH 14, 1977

The mother of my closest and dearest friend has shown increasing deterioration of Huntington's disease over the last ten years. Her experience is painful to follow. The hardest aspect of the disease from my perspective, however, is its effect on her offspring. Their lives are shadowed by a perpetual cloud of never knowing if it is safe for them to marry and bear children and share the life pattern that most take for granted. I hope above all that some kind of early detection research can be sponsored so that offspring of parents affected with Huntington's will know earlier on in their lives whether or not they have been carriers of the dreadful Huntington's syndrome.

Denver, Colorado

April 19, 1977

GRACE HERSETH
PHOENIX, ARIZONA

FEBRUARY 22, 1977

I work as a volunteer for Huntington's disease. The lady I work for, Lois Reed, devotes most all her time toward helping these people. I haven't had much contact with the really sick people, except for the two young women I know. It's a horrible thing to have.

This one young lady, in her twenties, has lost her children because of her inability to take care of them. It's a heartbreaking horrible illness, and I wish I could do more. If they just had a chance of getting well. How awful it must be to know you will die from it. I feel sick just watching them suffer. They are hard for me to understand as they don't speak clearly, and the one girl that lost her children is very quiet and lonely.

Anything that can be done to help them would be such a wonderful thing. We who are healthy can't imagine the "hell" they must go through.

Thank you.

Denver, Colorado

April 19, 1977

D. L. HOMESTEAD
LAKEWOOD, COLORADO

APRIL 1, 1977

I am writing to you about my concern for families with Huntington's disease and the Commission's meeting in Denver on April 19, 1977.

My mother has Huntington's disease and she had a sister who died of the disease 12 years ago.

I am married and have three children ages 20, 19 and 14. My brother is also married and has three children ages 16, 13, and 6. At this point in time, my brother, his children, myself and my children each have a 50 percent chance of being struck down with this disease. Conceivably, we could all have it or pray God, none of us. Had my brother or myself known of the disease when we married we could have chosen not to have children, limiting the possible carriers to two, not eight, as we have now. The information was not available to us, and, as a matter of fact, even today the limited information has to be sought out and probably many persons that are potential carriers of the "bad gene" are as unaware of the fact as we were.

Two of my children are of marriageable age. Soon they will want to know for certain if they will be able to have natural children without the possibility of Huntington's. I want them to have this information; my brother also wants to be in a position to counsel his family. Though we are willing to take a test that will give us the answer, there are no tests now available that can conclusively determine whether we are carrying the "bad gene." We need Federal funds for research to obtain this information and more than that, to find a cure for the disease itself.

There are also special problems in the care of persons having Huntington's that cause grave financial burdens on the immediate family, and some kind of Federal assistance is necessary. Perhaps it could be covered under Medicare or Medicaid for it is absolutely certain that sometime in the life of a Huntington's patient, total nursing care will be required. Because of the slow progression of the disease, this could easily average two to three years total care per patient. At \$75.00 per day, this is over \$6,000 per year, so you can see how desperately financial assistance is needed.

Denver, Colorado

April 19, 1977

I plan to attend the April 19th hearing, and if I can be of any further help or supply additional information, I will be happy to do so.

Denver, Colorado

April 19, 1977

THE REV. WAYNE V. HOUSER
BOULDER, COLORADO

MARCH 10, 1977

My interest in writing to you arises out of a pastoral relationship with one victim of Huntington's Disease. The patient died after several years of decreasing mobility. I watched the person slowly weaken until he needed full-time nursing care. His mother is a member of our church.

Though I am not informed about the medical and genetic aspects of Huntington's Disease, I am concerned with the emotional and psychological effects of the disease. I observed, especially in the last stages of the illness, a growing sense of loneliness and separation. This, of course, is common to all terminal illnesses. However, the inability to communicate adds to the problem.

Several of us, in order to try to help, researched the possibility of using a bio feed-back mechanism tied to a computer. This arrangement would allow minimal muscular effort to register messages via the computer to a cathode tube for reception by those in attendance.

We found that all of the technology is already existent. But funds are needed to adapt it and make it available to patients. I think it would be a great help in reducing loneliness and frustration.

Thank you for the work you are doing on behalf of those who are afflicted by Huntington's Disease.

Denver, Colorado

April 19, 1977

MAURINE JAMES
GLENDALE, ARIZONA

APRIL 19, 1977

How can one cover 20 years of life ravaged by H.D. in a letter short enough to hold interest?

My husband, and father of our two scns, succumbed to the ravages of H.D. in December 1968. The last six weeks of his life were spent in Arizona State Hospital, where he received excellent care. I am sure they did some experimental ministering. There are also movies of him at Barrows Neurological Center in Phoenix, where his condition was diagnosed.

I am sure education in this area needs to be available. Nurses at Barrows looked on in wonderment as tests were taken. I am sure they had never seen a patient with H.D. The rest home where I left my husband for 1-1/2 years in the daytime knew nothing of the disease.

After a thorough examination was made and the diagnosis came of H.D., nothing could be done except tranquilizers. We were given a handful of samples and were left to find the one that worked best. We finally stumbled onto one on our own. A chiropractor in Phoenix gave us counseling and a prognosis that helped more than anything. By the time we found him, H.D. had gotten to the last stages.

My husband's two sisters died of H.D. in Texas hospitals. His brother died in a government hospital in Waco, Texas, with H.D. One sister in this family escaped it. My two sons, now 37 and 35, of course are at risk. My four grandchildren also are at risk. The knowledge that research is being done along with a prayer on their behalf every day makes the waiting to see bearable.

Denver, Colorado

April 19, 1977

MARGARET T. JOHNSON, R.N.
BOULDER, COLORADO

MARCH 22, 1977

I have been told that your Commission is in the process of studying and making recommendations about the control of Huntington's disease and its consequences. I am a nurse who has helped to care for a young man who died a short time ago from Huntington's, and so am very grateful for the fact that there is increased interest in helping the victims of this disease, which causes untold suffering to them and to their families.

May I add my voice to many others who urge you to seek to provide more funds for research and also for help to those families who have the burden of years of care for their loved ones?

Denver, Colorado

April 19, 1977

NADINE S. JOHNSTON
LITTLETON, COLORADO

APRIL 5, 1977

I am 30 years of age and "at risk" for H.D. My father died of the disease in 1974, after years of suffering; but the suffering does not end when an H.D. victim dies. Each day I wonder whether or not I too will cause my family that much anguish. Will my personality gradually change to the extent that my loved ones and friends will not even remember the time when I was not fearful, angry, verbally and physically abusive and suicidal? Will I regret forever my decision not to have children? Will my husband be forced to put me into a nursing home, most probably one which is not educated or staffed for the H.D. patient? Will I make everyone around me miserable?

Each and every day of my life, I live with the memories and wonder what the future will bring.

Planning for the future may be futile for a person at-risk for H.D. However, my husband and I are extremely happy and are thinking of our future, planning the things most young couples plan, saving our money, investing, looking forward to retirement in our home state of Minnesota. Hopefully, all of our plans and dreams won't be wiped out by the medical bills associated with H.D.

I hope you will help me plan for the future by obtaining whatever help you can; financial, educational, and by putting most of the funds available to work finding a CURE for Huntington's disease. Yes, we need better care for the victims of H.D. Possibly this could be accomplished by educating the medical profession and the nursing care facilities already available.

If, through properly funded research, a cure could be found, the money directed toward new facilities could be saved. There would be no more Huntington's disease, which for me and millions of other people in H.D. families would be the best news life could bring!

Please help us!

Denver, Colorado

April 19, 1977

DOROTHY JONES
PHOENIX, ARIZONA

APRIL 15, 1977

The frightening spectre of Huntington's Disease has been with me for more years than I care to remember. At the time my husband and I were married, I did not know that this genetic horror had been in his mother's side of the family and that it ultimately would affect her as well as her sisters. My mother-in-law died of HD when she was 62 years of age, which preceded my husband's demise of HD when he was 51.

There were two children, a girl and a boy, born to us. To date the daughter has not come down with the dread disease, although it is a matter of grave concern to her. However, my son has been ill since he was 35 years of age and he is now 52. He married after returning from World War II and a son was born to that union. Unfortunately, this marriage ended in divorce; and there has been very little contact with my grandson other than support payments by my son until the young man reached the age of adulthood.

During the early years of my son's illness, he lived with me a good part of the time. He was very undependable and found it hard to stay in one place--wanting to travel around. This was a source of real concern to me, as his gait was impaired making him subject to falling and to even being picked up by the police as being potentially a public inebriate.

Because his illness is genetic and not service-connected, he has had some hospitalization in VA hospitals; but for many years now he has been cared for in a nursing home in Colorado. He is maintained by help from the VA, his Social Security, and welfare payments. For the past three years he has been unable to talk, which makes my infrequent visits to him seem almost to no avail. The last time I was there he kept wandering out of the room because, of course, we could carry on no conversation.

My greatest concern is that real effort be made to encourage research to find some way of halting or making less difficult this disease. I realize that much research is now going on and only pray that more resources may be directed into this effort. It may be too late to help my son, but perhaps there is hope for future generations.

Denver, Colorado

April 19, 1977

EDNA M. JOYNES
MESA, ARIZONA

NOVEMBER 8, 1976

This will be a brief outline of how Huntington's disease has broken up our family and caused us all untold sorrow and grief and has practically wiped out the life's savings of some. And to think there are at least 25-30 siblings who have a 50-50 chance of going through the same hardships and heartaches, if we don't find some help financially and--hopefully--a cure through research (all of which takes money and families involved to state the needs this dread disease requires in order to give our loved ones the care and attention they should have and courage to face the future--that it is not the beginning of the end).

My sister died last year with a stroke, after having the disease for six to ten years, the last two in a nursing home; and they had to sell their home in order to pay the bills. She even had a good pension, as she had worked 25 years as an R.N. with the State of Connecticut. I do feel there should be some kind of help in cases like these, whereby the family could get insurance or help before they have to spend their life savings.

I have two brothers living in Canada with the disease, both in nursing homes and getting excellent care. All the homes require from them for their care is their old age pensions and social security. The families are not obligated to pay any excess charges toward their care. It seems this great country should be able to take care of the ill and afflicted as well, if not better, than Canada.

My mother and her father, we feel, had this disease but it seems this generation is much worse than either my grandfather or mother was with it. My mother was worse with the involuntary movements than my grandfather. There were seven in my mother's family and she was the only one who had it. Four in our family, and so far, I am the only one with no symptoms and I am 63, the youngest in the family.

I trust we may soon be able to give the children hope and encouragement, if they do, or do not, have this gene that there is help on the way and that we are doing all we can to control or cure it, to enable them to lead normal, useful lives.

Denver, Colorado

April 19, 1977

RICHARD E. KEENER
ARVADA, COLORADO

APRIL 18, 1977

I was scheduled to testify before the Commission on April 19, 1977, however, my employment necessitated me being out-of-town on business that week. Therefore, I am writing this so it may be entered as my testimony.

My mother-in-law has Huntington's disease, and only in the past month has been admitted to a hospital probably never to return home. We now face the same problem that millions of others face daily--watching a loved one slowly waste away.

While that is bad enough, it by no means begins to express the real problems inherent in this dreaded disease. We went through several years of anguish as my wife and I watched her mother begin to stumble, slur her speech and exhibit other symptoms that no doctor could explain. At one point, she was committed to a mental institution due to the outbreaks of violent temper. Again, she was diagnosed improperly. Only after one of her sisters was diagnosed as a victim of Huntington's was the real problem understood.

My wife now faces the 50-50 chance faced by all children of the Huntington's victims. She has the knowledge that some day she may be exhibiting the same actions she finds so depressing in her mother. The effect this has on a person is something you have to experience to understand.

I make this my plea to the Commission to do as much as possible to aid those stricken with this and other such hereditary diseases. At a time when the President's wife is in charge of National Mental Health, it would certainly be fitting that a great assault be made on Huntington's disease.

Assistance is needed not only to further research for prevention and cure of Huntington's disease, but to simply create a worldwide understanding of the disease itself. Countless thousands have watched a loved one deteriorate away in an asylum without ever knowing the true cause.

Help is desperately needed in the form of assistance and counseling for those trying to care for a Huntington's victim.

Help is also needed for the children of those stricken with H.D. as they try to build on a future they desperately fear.

When billions of "our" tax dollars are sent to causes all over the world, it seems like a simple portion of this be provided to give "hope" where none now exists.

Denver, Colorado

April 19, 1977

JACK A. KLAPPER, M.D.
DENVER, COLORADO

MARCH 15, 1977

Regarding your letter requesting information about the private practice experience with Huntington's chorea I would like to say the following. I see approximately one to two cases of Huntington's chorea in a year's period. Generally the advanced cases are managed just as chronic neurologic disease of other types is managed. Institutional care and nursing home care is frequently needed, and I find no difficulty in arranging this, and feel that this is the best available placement in many instances. The problem stems in dealing with the next generation. Young people often come to my office with a story of a family member or a parent having been told or diagnosed as having Huntington's chorea, and the inquiring patient then wants to know how she can find out whether or not she has the disease. This, of course, is a difficult area and requires extensive testing which is often not productive of definite answer. These people are often interested in learning more about the disease and are looking for academicians and researchers who might be able to help them. I would think this kind of service might be quite useful to these people. Genetic counseling, of course, is probably the only thing we have to offer these people at this time.

Denver, Colorado

April 19, 1977

PAMELA KNISS
DENVER, COLORADO

MARCH 30, 1977

It was not with a sense of jubilation or relief that I finally learned that the disease had probably not cursed my mother and that I was therefore free to have children and to pursue my professional goals. Because of the fear and helplessness suffered by my family and by others touched by Huntington's disease, it is now with sadness that I make this statement.

Needless fear and anxiety influenced my family's life for many years. The cause was ignorance and an unwillingness by family members to speak of the mysterious illness that afflicted my grandmother and her three sisters and that shamed the family. Even the doctors involved in diagnosing my grandmother's disease, did not give my mother specific details. She would not learn of these details until years later, when they were mentioned casually in a college lecture. Left stunned and panicked, my mother sought the help of another doctor. He assured my mother she had nothing to be concerned about, however, he gave no details and would not meet her eyes. Because of the lack of honesty concerning the disease, my mother became so anxious about the future that suicide seemed the only alternative. Her death would have protected my brother and I from knowing the same fear and helplessness that she experienced daily.

As a child, I can remember sensing that my grandmother was not well. I became irrationally afraid of sickness and death and was always afraid that I would "catch" my grandmother's illness. I also blamed myself for causing the periods of anger and depression experienced by my mother. My grandmother lived near or with us for ten years. She placed an unbelievable strain on a marriage that subsequently ended. Because my mother was my only parent, I became afraid that she also would die and leave my brother and I alone. Fear was accompanied by anger at having to constantly confront the irascible nature of my grandmother. This was replaced by guilt when institutionalization was the only solution to her care.

Finally, as a result of a story dramatized on Marcus Welby, a television program, I became aware of the terrible ramifications of Huntington's disease. I sought genetic counseling at the University of Colorado Medical Center and

It was because of this experience that my family could at last fully understand the disease. The doctors gave us no false reassurance. Through honesty, they allowed us to assess our own situation and to then make decisions concerning ourselves and our families' futures.

The fear and suffering caused by ignorance is needless. Education is the only means of helping individuals and families to deal realistically with their problems.

In order to meet the needs of the public effectively and efficiently, dissemination of material concerning specific information about the disease and available services must be coordinated with the activities of existing agencies. Pamphlets distributed by agencies like Planned Parenthood and placed in doctors' offices, family planning clinics and waiting areas of other health facilities would reach many people. The Government Printing Office may also provide pamphlets for public use. The Department of Health, Education and Welfare could use its political influence to initiate educational activities for personnel in university medical schools, college health facilities and public hospitals and schools. It is also important that individuals in the fields of mental health and alcohol and drug addiction treatment be given information, because many families seeking help for an afflicted family member view the problem as only one relating to mental health. Finally, advertising on television and in popular magazines will inform large numbers of people. The anxiety, anger, guilt and great sadness associated with Huntington's disease will never be eliminated. However, individuals and families deserve the information and help necessary to make reasonable and responsible decisions concerning care and treatment, family planning, and resources available for helping those left to cope with a family member's illness.

Thank you for this opportunity to share my thoughts and thank you for demonstrating concern for those touched by this disease.

EDWARD J. KRAMER
PHOENIX, ARIZONA

APRIL 12, 1977

I will give a resume of my situation and then follow up with my views on care, particularly of patients with advanced symptoms.

My first wife, I know now, suffered from H.D. We were separated, then divorced. I had custody of our two daughters, Edna and Kathie, from the time they were two and four years old. Their mother died in 1942; the children were then 7 and 9 years old. I remarried in 1943. My second wife died of cancer in 1960 and I have not married again.

By 1954 my younger daughter, Kathie, showed signs of emotional disturbance and received private psychiatric treatment. After some private hospitalization she was ultimately diagnosed as schizophrenic and entered the State Hospital. Not too long later she was released and spent a semester in college, married and gave birth to a son in 1957. Her condition rapidly worsened and she was again admitted to the State Hospital where she remained most of the time until her death July 19, 1970, at the age of 34. Autopsy report: "Severe bronchopneumonia immediate cause of death, and a general condition of emaciation and dehydration. No genetic correlations." (Kathie's son will be 20 years old in May, close to me in family relationship but lives now in neighboring New Mexico. No H.D. symptoms.)

During Kathie's hospitalization Edna, after a successful secretarial career, showed signs of mental problems in 1967. She was in several private institutions but eventually broke down completely and entered the State Hospital in December, 1968 on a voluntary admission. The diagnosis at this point was "schizophrenic." After Kathie's death in 1970, doctors at the State Hospital said that both Kathie and Edna had H.D.

Edna now requires total care. She cannot speak, walk nor care for herself in any way. As I am retired (since 1972) I am able to visit her daily, taking special foods to supplement her baby food diet.

As outlined, both of the girls showed mental disturbance first, as did their mother. Kathie later developed a muscular rigidity while Edna now has pronounced chorea movements.

Edna is in the Medical Division of the hospital and receives the best care possible with the available equipment, all it seems that is available anywhere. It appears to me that an important area of research which would help H.D. patients NOW would be in the field of care equipment. Much could be done with wheelchairs, for instance. They could be large enough and sturdy enough for the hyperactivity of the patient but at the same time designed to avoid bruises, cuts and scratches. Now the nurses' aides tape padding over many areas of the chair, trying to avoid such injuries, not always with success. The manufacturers seem to skip from the normally constructed wheelchair to the geriatric chair, which just does not do the job for the hyperactive patient. Also, where a variety of restraints are often required, these could be planned to give the patient more freedom with safety. Too often the aides have to rely upon equipment made with restraint as the top priority. This is also true of the Posey belts used both on beds and chairs, most of which have locking metal buckles. Belts not using the metal buckles require special instructions so are not often used. The beds themselves could be greatly improved for the comfort and safety of the H.D. patient. Sometimes extra mattresses are put on the sides of the bed, and sheepskins on the headboard. They don't always stay in place. Incontinent diapers and pads are often inadequate for the frustrated, humiliated H.D. patient. Perhaps a problem in the training of care personnel arises because H.D. patients are usually scattered in various wards and nursing homes. If H.D. patients were more centrally located, it might help greatly in both care and treatment.

Having already been so critical, I might as well add a personal viewpoint on discussion of the hereditary nature of H.D. Out of well-meant compassion, many doctors and other professionals tend to side-step this problem. Until definite research progress has been made, I feel that H.D. families should be given more realistic counseling.

I will not be able to attend any of the Commission hearings but you can use the contents of this letter as you see fit.

BRENDA KRONBORG
MESA, ARIZONA

NOVEMBER 5, 1976

My name is Brenda Kronborg and I'm 16 years old. I'm writing this letter to tell of my experience of Huntington's disease. This dreaded disease has hung over me all my life. When I was younger, I asked my parents about my mother's grandfather. He had died before I was born. My mother never deceived me and told me the truth. My great-grandfather had a disease thought to be Parkinson's. The only thing they knew at the time, 1958 or so, to do was to confine these people to institutions hoping that this might help. He was confined to an institution in Chlorenda, Iowa. He would come home occasionally and it was believed that on one of these visits he found a letter granting permission for my great-grandpa to be permanently confined to this institution. He was found by my uncle in a neighbor's garage on the same weekend that the letter had been signed. He had hung himself. His mother had the same disease as he had and my grandmother has. I used to tell my friends what a great grandma I had.

I moved from Iowa to Arizona when I was 8. We used to fly back for Christmas and New Years (until my mother started working for a company that has its busiest time of year at Christmas). My grandma would have all kinds of candy, nuts, cookies and meats just because it was Christmas. She'd sew me clothes for any occasion or reason; and grandma's clothes would always fit right. One of the best things about her was that she could easily pass for my mother. My mother and brothers and I drove to Iowa when I was 12 in the summertime. It was then that I noticed a difference in her. She was 52 then. Her hair had grayed amazingly; her face had wrinkles; and she seemed tired, but yet she could never seem to sit still, and most noticeable--her legs were wobbly. Her legs always seemed like they were going to give out; she had to grasp the railing when she went down the stairs. I just thought that my young grandmother had grown old and pressures were weighing her down. My father couldn't make it to Iowa that summer, but he saw my grandparents the next summer. My dad was amazed at my grandmother. She slipped and dropped things all the time, her speech was slurred, and her body was constantly trembling. He told my grandpa of a good doctor who could refer him to a specialist for my grandma. Since then, she has had many tests and tried different medicines in the attempt to help fight Huntington's. I hope someone will come to find a cure for this before my Mom's time to have it comes (if it comes).

Denver, Colorado

April 19, 1977

MAE KRONBORG
MESA, ARIZONA

NOVEMBER 5, 1976

I am Mae Kronborg, 38 years old. I have seen Huntington's in three generations of our family and it doesn't give a person a very secure feeling about it. My great-grandmother died of it in 1957. We didn't know what she had; us kids were told she had Parkinson's disease. When grandfather was around 50 years old, he started dropping things; not nearly as sure footed as he had been. He was sent to a mental institution which broke him down in every aspect of life--tied his hands and everything else. He came home, found a rope, and hung himself in 1960. My grandmother did find out it was Huntington's. He had one brother that also has it.

All three of his children have it: my mother, my aunt and uncle. My mother has known for about a year now. When we found out it nearly tore us apart. She doesn't really have any good doctors in Iowa that know about it. The medication she was taking, she couldn't take. When she was on it, she couldn't talk. She has lost a lot of weight and she needs constant help.

It would be so very nice if we could get help for all these loved ones and know that when my time comes, if I have to have it, I will have help for it.

SANDRA LAMBERT
PHOENIX, ARIZONA

FEBRUARY 10, 1977

I am writing to the Commission because I have Huntington's chorea.

I have had Huntington's five years, so I'm truly interested in the Commission. During the five years I've had trouble in walking--even like writing is difficult for me, because I shake so much that holding my pen is hard for me!

And things like eating is difficult because of swallowing is hard. I have trouble with urine. All these are difficult just because I have Huntington's chorea. I inherited this from my mom who people thought was an alcoholic all of the years until she died.

None of my family visits me ever, since I told them that I had H.D. like Mom had. I took care of her for three years before she died. I was the only one in the whole family who cared for her. They treated both of us like we had the plague! In the end I fed her baby food because that's all she could swallow without choking.

There were times when depression set in that I wanted to end it all. At the time I thought I didn't want to be treated the way they did my mom. No one even came to her funeral. But now I have learned to live each day as it comes, knowing what I know now about my illness.

Each day I live in hope--that there will be hope for me and my kids; they took them because of my illness. All of them are in foster homes. I now can live in hope for a cure with the Commission!

You can print my name.

MRS. MARTHA (ENNS) LEMEN
PUEBLO, COLORADO

APRIL 4, 1977

The problems facing families with Huntington's disease are many, the foremost being unable to find professional help knowledgeable enough about the disease to be able to assist patients and families, and facilities to care for them.

My mother's illness was diagnosed as Huntington's disease at a time when it was too late for me and my five sisters and two brothers to make a decision as to whether to have families. Being an at-risk person I felt indebted to my husband and family to do what I could to get an early diagnosis so the disease could be stopped. A feeling of guilt and helplessness not to be able to advise our two children concerning marriage and families of their own until I passed the at-risk age.

My childhood memories of our home life are those of continuous conflict between parents due to my mother being unable to cope with day-to-day living and problems that normally arise in rearing a family. Her main concern was who would care for her and her family as she slowly became physically more disabled day by day, a process that went on for 15 years. The last five years she was bedridden requiring total care.

Costs of hospital care for such a lengthy stay are prohibitive; nursing homes and mental hospitals don't have enough personnel to take care of these patients. Family members are forced to stay employed to cover the many medical expenses, so education for the children must be postponed to enable them to help with duties at home.

Since my mother's death, one brother and two sisters have been diagnosed as Huntington's disease. I'm wondering where, when, or how will it ever end?? It's like an epidemic that won't go away. Who's next? Something has to be done, somehow!

These are only a few of the problems the victims and their families must face: Families are unable to help each other because of distance. Families are being moved because of employment.

Help for housekeeping is unavailable and hospital care prohibitive for such a long, terminal illness.

The patient is aware from past history, of the prognosis of the disease, which causes severe mental depression and suicidal tendencies.

The first and early symptoms of Huntington's disease are the unexplainable, gradual personality changes that occur and are difficult for families to accept.

The choreic movements are so similar to an alcoholic; their lack of coordination makes it difficult to take them out in public.

So much time and money has been spent in search for physicians who have enough knowledge of the disease and who are able to diagnose or treat it.

Their advice in most cases is to admit them to a mental institution for care as a terminal patient.

I sincerely hope this testimony will serve as a means of getting help to thousands of other families with similar experiences. Possibly it's too late for our generation but we must do what we can to save our younger generation.

Denver, Colorado

April 19, 1977

JUDY MATOUSEK
PHOENIX, ARIZONA

FEBRUARY 26, 1977

Please forgive me for making this such a long letter. I wanted very much just to write Huntington's disease victims is a living death, HELP! But as I started writing, I realized that I was begging for help for people, not a place or thing you find in the store. What I am asking for is a need for care and help for these victims now and in the future.

Years ago there was this beautiful lady with long, auburn hair, blue smiling eyes, sweet and kind in her words and actions. She worked during the week to make ends meet, with raising two small girls and her brother. It was a must. Her father had died when she was younger and her mother was in a state hospital.

She was the lady who drove the kids to the show or skating rink or wherever they wanted to go. This same lady made her girls' clothes on her days off. The same lady who took her vacation time to take the Girl Scouts camping in the summer. On the weekends she would never miss going to see her mother. I always wondered why her mother never got well and came home. She had been in the hospital for the past eight years.

Well, every night this lady would go into her little girls' room to kiss them good-night and they would say their prayers together. One night one of those little girls asked why the doctors didn't make Grandma well, so she would come home. And this beautiful lady looked at her with very sad eyes and said, "The doctors didn't know how to make Grandma better, but maybe God would soon take her to Heaven." She now had tears in those eyes and I wanted to cry for her too. I never dreamed that some day I would have those very same tears in my eyes when I talked about her; you see, that lady is my mother.

All those years that she was raising us I thought all mothers did the things she did for us. But that's not true; she indeed was a very special mother. She tried to do all the things she could for my sister and me while she could. She knew that someday she might have what her mother had--Huntington's disease.

I'm omitting a few years now because they were hard and long years for our family; mother was changing, and none of us knew why.

During the past 15 years we watched this beautiful, giving person become a NOTHING HUMAN. Just a thing that eats and breathes, no pride, no giving love, eyes expressionless, unable to stand or walk, unable to hold her body still. When I look at her, I wish I could shout out NO, NO, NO; if only I could put down on paper the things in my heart and mind. Oh, how I pray that she would be the last.

It's a summer day, but I've got to be asleep because it has got to be a nightmare. This really couldn't be happening to me, not in this day and age. Sitting on the side of me is my father and on the other side is my husband; it's very quiet in the room. It's old and bare and hot. There are two flags in front of the judge's desk; standing in the doorway were two men talking about something. The judge was reading something on his desk; behind him on the wall was a clock reading 10:00 o'clock. One of the men walked over by the desk and began reading a piece of paper; I heard him say my mother's name. I don't recall all the words he said, except that Madelene Karl was completely incompetent, a burden to her family and dangerous to herself. Was I really hearing these words? They seemed so evil, like in the dark ages; this is the 20th century. My God, what are they going to do with my mother? The judge is talking now, "It is the recommendation of this court to have MADELENE V. KARL committed to the state hospital for the insane, until such a time she could return to society without being dangerous." Oh, my God, what have we done? There is no return, there will never be such a time. Why can't I say something? Don't just sit here. Tell him he's wrong. She is not insane--she is sick. She has Huntington's disease. Those tears come now, the ones she had on that night I asked if the doctors could make Grandma well. Never had I had tears like that; would they ever stop? As we walk outside, it was hot and I thought I've just been to my own mother's funeral--only she wasn't dead. My tears did stop and they don't come very often now, just deep scars in my heart. You see, that day in court was almost twelve years ago.

This is not the end of my story; it's only the beginning. You see, I have two little girls, Kathy and Misty. What if I'm the next H.D. victim? Will they, too, have to sit in a courtroom one day? Oh, my God, I pray not. And that's where you come in.

Denver, Colorado

April 19, 1977

Please help all of us. It's people like you that can give us the help we need. A place where we can have help, care, love, understanding, medicine, and maybe even a cure. Please help! Oh, I never did tell you this--that day in court, my mother was only 38 years old.

Denver, Colorado

April 19, 1977

SPENCER I. MEIER
AURORA, COLORADO

APRIL 2, 1977

I am writing you regarding my views on both the tragic implications of Huntington's and the needs of the thousands of individual human beings whose lives are affected by it. Surely, by now, the Commission has obtained a fairly comprehensive picture of the very real human tragedy associated with the disease. However, there are other pictures to be painted. There is an ever brightening picture of hope, and this picture can be painted even brighter by the actions we take right now.

First, you must grasp the scope of Huntington's. On the Colorado CCHD mailing list there are about 200 names. It must be realized that these 200 names represent at least ten times that number of people affected, just in this area. You must also realize that the very small percentage of us who are writing is not indicative of the percentage who care. The majority are, understandably, reluctant to write and, likely, afraid to think about it.

To grasp the scope of Huntington's on the personal level, I will relate to what is represented by just one of the names on the mailing list. This name represents an old woman, widowed 15 years ago by Huntington's. She has two children at risk, and beyond that, six grandchildren. She is emotionally crippled by her memories, fears, and guilt. Through her eyes there is nothing to look forward to but to see the tragic cycle of Huntington's rape her life once again. She takes no joy from life. She wants to die.

The mailing list represents not only persons diagnosed with Huntington's, but those at-risk and all family members whose lives are influenced just as surely as if they had the disease themselves. For those at-risk it's a waiting game--a game they eventually lose no matter the outcome. Should they ignore it? Should they be ashamed? Should they have children? Should they design their whole lives around the possibility they will be stricken? The statistics of Huntington's not only mean a 50 percent chance of getting the disease, but also imply a very important 50 percent chance of not getting it. But how do you deal on a personal level with those figures? How do you live a 50 percent normal life? The answer is you can't, and the point is that Huntington's affects at the rate of 100 percent those lives of persons at-risk and their

families. Something must be done and there are no real solutions short of a control or cure for this most dreadful disease.

What are the answers to Huntington's? Is there hope? I believe there are answers and there is hope. However, this depends completely upon what we do today, upon the actions of your committee, and upon the continuing active work of CCHD and people like Marge Guthrie. It depends upon immediately increasing the levels of medical research and treatment directed toward a cure. I can look into both the past and the future. I see that Huntington's must be a truly ancient affliction because it is spread to all races in all parts of the globe. In even the recent past Huntington's was thought to be so rare that it could be safely ignored--leaving entire families to face the specter of H.D. alone, and without hope. This situation has drastically changed and Huntington's has been brought to the attention of society's moral and medical conscience. I see that medical science has finally advanced to the point it can provide answers. I see a day in the future when Huntington's will be controlled by something maybe as simple as a dietary matter. This day will come in within five years or 100 years, and that time schedule, to be measured in terms of human suffering, is largely up to what we do today.

In addition to the all-important need of funding for medical research, funding is needed in the areas of treatment and education. I also see a need for more members of the medical community to devote their work to Huntington's.

In all too many instances Huntington's is a sideline of a few physicians or, even worse, H.D. patients are treated by those who know nothing of the problem.

A unified approach is needed. In large metropolitan cities, centers could be designated which are devoted to the treatment and care of H.D. patients in the area. Huntington's must be realistically viewed as something which must be dealt with not only for those who have developed symptoms, but for every person associated with the disease. Therefore, these centers should also provide counseling and educational services, hopefully, on the most optimistic tone possible. In a few of the larger centers, active medical research could be carried out by interested professionals.

The educational thrust must be directed also at removing Huntington's from the category of a "closet disease." Education and counseling may be able to loosen some of the social stigma associated with the disease, and give those families involved enough fortitude to help participate in the fight for its ultimate cure.

I wish to thank the members of the Commission for caring. That's what it's all about. I am also grateful for the opportunity to do my share in this noble cause.

Denver, Colorado

April 19, 1977

MRS. KENNETH MONTEL
FORT MORGAN, COLORADO

FEBRUARY 23, 1977

I am not a member of the immediate family nor am I an at-risk person of Huntington's disease but I am a sister of one who has suffered through the rigors of Huntington's disease with her husband and her young son. Because of this, I wish to emphasize to the committee the necessity of immediate concern and funding for the research, promoting the cure of this disease.

My sister, Dorothy Johnson Snyder, of Boulder, Colorado, has seen the disease come into her life, not knowing anything of its magnitude. She inquired and searched through medical channels and her own research until she knew what was ahead for her. Through the knowing of its complications and the hope of future cures she faced her husband's illness, death and then the awful realization that her twenty-year-old son was a victim. The love and compassion shown him as he, nine years later, chose to starve himself in order to more quickly end his life, is a sainthood experience that few Huntington's family members can claim. Only by faith that a cure might be found soon could she have so beautifully come through this experience. It has left scars on her life but with the hope of positive research she continues eagerly to help this cause. But this can only be done by your committee's action.

With this having so closely touched my life, I feel qualified to urge you to do all you possibly can, as a committee and individually, to promote and work diligently for more funding for research for this disease.

Denver, Colorado

April 19, 1977

JEAN MORLEY
PHOENIX, ARIZONA

FEBRUARY 24, 1977

My husband is at risk for H.D. We found out about five years ago that his mother has H.D.

We had never heard of H.D. so we did all possible to find out more about it.

We were very shocked when we learned it is an hereditary disease. We have two sons and it hurt to realize that this disease might be passed down to them.

So far my husband has shown no signs of the disease; he is 40. We worried about what the company he works for would do if they found out. They did find out last June when my husband had his yearly physical for the company. It seemed to make no difference so we were very relieved.

I know my husband worries about getting H.D. but he doesn't dwell on it. We live each day as it comes and just pray that God will give us the strength to cope if he does get it.

We tend to worry more about our boys. Our youngest has some nervous habits--eye blinking, etc., and we wondered if it could possibly be H.D. The fear is always there even though we're not always conscious of it.

My husband has two sisters--neither of them really care to discuss it and one will hardly admit the disease exists. My husband and I believe in facing problems head-on and feel that what we don't know will hurt us.

My husband's parents are living in our little guest house. Dad retired last August and they moved here from Colorado so that we could be close and help care for Mother. Her medication has really helped control her movements and she has done well. She is getting worse and we know there will be hard times ahead. But we are so grateful she has done so well for so long.

We do what we can to support our local CCHD Chapter. We do realize how great peoples' needs are. Research is at the head of the list. Better care facilities, home nursing help and a better understanding of H.D. in general are needed.

Denver, Colorado

April 19, 1977

One of our hopes is that there will be a test to find out if the at-risk person is going to have H.D. Then we could give our children the facts and they could have a choice as to whether they will have children.

Denver, Colorado

April 19, 1977

MR. AND MRS. RICHARD MORLEY
PHOENIX, ARIZONA

FEBRUARY 23, 1977

I am writing in behalf of my wife and three children. When we married we knew nothing of this terrible disease, Huntington's. My wife got sick and I took her to several doctors and finally our doctor confirmed she had Huntington's disease. By this time we had had two daughters and one son and they had eight children among them. Now these three children are living in fear they might get the disease and if they get the disease, their children have a chance of taking the disease also. In my judgment, there is a need for more and improved research and training to lick this disease. All we ask is that you do your very best. Thank you.

ELSIE L. MURPHY, R.N., M.S.N.
MESA, ARIZONA

MARCH 25, 1977

As a registered nurse for twenty years I have seen several patients with Huntington's disease. The first was when I was still a student in Kansas. The lady I saw there made a great impression upon me. She was in the final stages of her illness and in a locked psychiatric unit--a frightening and grotesque spectacle to a young student. This woman's daughter was just being diagnosed as also having Huntington's disease.

Throughout the years I have seen other Huntington's disease patients in various stages of illness. For the past five years I have served as nursing advisor to the Arizona Chapter of CCHD. In this capacity I have observed the potential Huntington's disease patients, persons with Huntington's disease, and the families of these persons.

During these five years, I have observed the difficulty families have had finding adequate help and care for patients after they are unable to care for themselves, and lack of knowledge of Huntington's disease by health care professionals. However, by far the greatest interest and concern I have observed by patients and families is in regard to research and the hope that a cure or a prevention will be found which will be available to children and grandchildren not yet afflicted. There is a great need for assistance in all three of these areas.

KATHY O'NEILL
DENVER, COLORADO

FEBRUARY 9, 1977

This is in regard to your request on suggestions and special problems of HD patients and at risks.

I am an at risk, my mother has HD and both my parents live out of town so I don't see a lot of the problems that I would if I lived at home. Some of the problems though that I noticed are as follows:

1. My Dad had trouble locating a doctor for my Mom.
2. It took a long time to diagnose it.
3. Right now my Dad and brothers have to take over most of the cleaning, cooking, and regular chores at home.
4. I have a 7 year old sister who has seen some of the violence that accompanies this disease.
5. It seemed to take a long time before they found any medicine that would help calm my Mom down. (I don't know if this is normal but I imagine it is.)
6. I am 26 and I would like to know how far this disease has to go before you should go to a doctor to diagnose it. (As you all probably know any little thing that is normal to other people such as twitches, clumsiness, etc. make me wonder if I am next. Also where to go to diagnose it. We (my family and I) have been down for tests at the Medical Center but that seems to be the end of it.
7. I have questions about adoption. I haven't even inquired from adoption agencies because it also seems like a bad idea after I see what my little sister is going through.
8. I would like to do more than just contribute to this cause, but after I saw the movie and the facts were brought out, I realize how bad it is and it only keeps me from sleeping at night, to see some of the patients etc. etc. etc.
9. If they find a test to see if you carry the gene or not will they tell the patient at risk. Or will they wait until he requests it or not give it out at all until they find some controlling medicine? I feel that they are getting nowhere with this. How long have they actually been really working on this?

I don't know if this will help but I hope so.

Denver, Colorado

April 19, 1977

SUSAN PETERS
THORNTON, COLORADO

MARCH 21, 1977

My name is Susan Peters. My mother is a victim of H.D.

It is very hard for me to write this letter for there are so many things that have happened with my mother as I stand by helplessly watching her die.

My mother was a very outgoing person, straight A student with a college degree, loved to bake her own bread, and loved her garden; and now she weighs 50 lbs., is unable to talk, walk, or do anything but lie in bed staring out of the padded sides of her living prison.

The first thing I would like to talk about is the guilt my sister, brother, and I feel now because of the first stages of H.D. The hatred the three of us had against my mother (there were many times when I called her a witch). She would turn on us so violently for no reason at all and we could not understand why, because she was always easy to get along with. We just all thought she was crazy and hated her with a passion, for she appeared normal to us except for these outbursts of anger. Now we all regret the many painful words we yelled at her. If we had only known she was sick, many unpleasant memories would not haunt us now.

As far back as I can remember I have always wanted two children (lest we only have one). When my mother's disease was diagnosed as H.D. my husband had a vasectomy. The only reason we have our son is because it took the doctors five years to tell us what my mother had because so little was known about H.D. Why? Why is so little known that my mother wasted five valuable years of time and proper medication searching for what was wrong. By the time it was diagnosed as H.D. it was already too late for her and our life of hell was just beginning.

You may ask yourself why did I say, "a life of hell?" Well, I will just tell you of our most recent episode. In the summer of '76 someone on the nursing staff at Plaza Care Center gave my mother a triple overdose of thorazine. Her heart stopped on the way to the hospital and the doctor in emergency told us she only had a short time to live; but she made it. We contacted a lawyer but our hands were tied because the doctor

backed down and wouldn't testify, and the hospital would not release her medical records. So the murder attempt, as the doctor called it, goes by unpunished and unnoticed by all. Why? Why should things like this happen?

In closing all I have to say is I hope research is funded so many questions can be answered on H.D. So that if in the future I get this disease I will not have to kill myself to spare my husband and son. The hell of life they would have to bear with me around would be unbearable for me.

Denver, Colorado

April 19, 1977

JEANNINE M. PIRES, R.N., M.S.N.
PHOENIX, ARIZONA

MARCH 31, 1977

I am a registered nurse responsible for total management of the 140 bed Geriatric Unit at the Arizona State Hospital. The majority of our patients are 65 or older, but we do also care for younger persons who have severe organicity affecting their behavior. Many of our patients are not appropriate for nursing homes due to their behavior management problems.

Among our patients we currently have two diagnosed as Huntington's disease, and one who is right now undergoing a complete diagnostic work-up. The extreme physical disabilities suffered by the Huntington's patient make program management for them very difficult. We group our patients on wards according to their ability to physically care for themselves. Our two patients with Huntington's disease have a good ability to comprehend their environment so we prefer to keep them on a ward with more social patients. Their physical disabilities to care for themselves makes them less able to be as involved in programs as others in their living area. It also places an extra burden on staff time. They need a more stimulating environment than they can get in our total care wards.

One of the two patients was able to go recently to the special clinic operated by Dr. Stern at the University of Arizona in Tucson and on return has done considerably better. The research, evaluation and new techniques there did seem to afford him some improvement. His transfer there probably would not have occurred without the interest and encouragement of his wife. We need more such clinics and more conveniently located so that persons with Huntington's disease can get the help they need and more research can be done. It really is tragic when these people have to spend years and possibly all their last years in a State Hospital when their problems have a very physical and not just mental origin. I would hope that additional funding can be approved to carry on the work for Huntington's disease already in process.

Denver, Colorado

April 19, 1977

THEODORE T. PUCK, Ph.D.
ELEANOR ROOSEVELT INSTITUTE
FOR CANCER RESEARCH
DENVER, COLORADO

MARCH 7, 1977

I know you are aware of my concern about the need for more support in investigation of Huntington's disease. I am writing this letter in hopes that it may help influence decisions of the HD Commission and assist in obtaining needed action in this field.

The enormous tragedy which this disease entails is of itself enough to require application of much more resources in an attempt to control its dire effect on human lives. The present time appears to be particularly propitious for renewed efforts in this area. The relatively young discipline of somatic cell genetics and genetic biochemistry has been developing impressive powers which now seem able to promise fundamental new understanding of the underlying defect in this disease. Such understanding might well provide methods of prenatal diagnosis, more effective genetic counseling, and ultimately, perhaps, a new and more powerful form of treatment. Moreover, the careful study of this disease by means of these new scientific approaches should yield information useful in understanding and ultimate control of a number of other related diseases which also still represent tragic enigmas.

It would be a great pleasure to show you some of the work of this laboratory when you are in Denver for the forthcoming commission meeting.

I send warmest regards.

ARTHUR ROBINSON, M.D.
NATIONAL JEWISH HOSPITAL AND RESEARCH CENTER
DENVER, COLORADO

APRIL 4, 1977

The following statement should be considered part of the official proceedings of the Commission, which I understand will be meeting in Denver, Colorado on April 19, 1977.

There are many unmet needs currently in families, one or more members of which are afflicted with Huntington's disease. One of the needs is the identification of those at risk for the condition and their counseling and support. The problem here is manyfold, particularly the education of the paramedical and medical professions in the principles of human genetics and the techniques of genetic counseling. This need, major as it is for Huntington's disease, may be generalized to a much more significant problem relating to all genetic disease in our society. The first line of defense is the family practitioner, who currently is woefully lacking in this general area. Much can be done today in genetic counseling and prevention of Huntington's disease, and even in alerting physicians to the early diagnosis of the condition itself. More should be done through the education of the child or young adult at risk, through support of the individuals in the early stages of the disease, and through the physical and mental care of individuals in the more advanced stages of the disease. All the above we are currently capable of doing. We have not made the necessary effort to provide medical and paramedical professions with the information or the personnel with which to carry these out.

An obvious need in research is in the general area of early diagnosis of those at risk of being affected prior to reproducing and even for the diagnosis of the "carrier" state in utero. Psychological and emotional implications of making this diagnosis early in life should be examined. Fundamental studies as well in molecular biological and biochemical approaches to the understanding of the primary lesion of this single gene defect would be of immense help, both in early diagnosis and in understanding of the fundamental processes of the condition.

I suggest that this Commission would attain its long-term goals more effectively by approaching the entire problem of research and services in human genetic disease, since this in the long run is likely to be more effective than the "disease of the month" approach. Similarly, the patient care problems are not necessarily unique to those with Huntington's disease, but are generalizable to most chronically ill adults suffering from neuro-degenerative conditions.

Denver, Colorado

April 19, 1977

CAROL SAWYER
MESA, ARIZONA

APRIL 22, 1977

My mother-in-law has Huntington's disease. She is one of ten children, and six of the ten have had it; four of them are dead from it. My mother-in-law didn't display the symptoms until she was in her fifties, and then the early symptoms were misinterpreted as those of menopause. It is very hard to see someone who you love and who has loved you become grotesquely ill, and even harder to see their own mental anguish as they see themselves becoming grotesque.

I married the oldest of her six children, and we have two daughters who are just entering their teens. And they have 16 cousins. So there are 24 people at risk in our family and the six spouses and their families whose children's and grandchildren's lives are affected by H.D. All of our lives have been torn apart emotionally, not only with dealing with those in the family who are already suffering from the disease, but with the limited prospects for our own futures.

We are hoping research will bring some relief for the worries about our futures, and we are doing what we can to support that research; but our resources are limited, and we know that something must be done in a bigger way to help the researchers find some answers for us. Please help us if you can.

Denver, Colorado

April 19, 1977

DOLORES SCHROFF
PHOENIX, ARIZONA

MARCH 3, 1977

I am writing on the behalf and for the millions of people that are afflicted with Huntington's disease.

This is a terrible dreadful disease that eventually destroys all of the body and mind and is very distorted while doing so, and puts thousands into state mental hospitals. Some of these people could be saved of some of this if there were enough funded to this cause so they could get the right medications, have visiting nurses and so on.

Won't you help?

Denver, Colorado

April 19, 1977

CARLA SEABOURN
TUCSON, ARIZONA

FEBRUARY 25, 1977

I am writing this letter, not because I am a Huntington's disease victim, but because I am so thankful that I'm not.

My father's mother died from Huntington's disease in a mental institution. Two of my father's brothers and one of his sisters died from Huntington's disease. Had my father developed Huntington's disease, I, too, would have a 50 percent chance of being a Huntington's disease victim. Had I developed Huntington's disease, my daughter would also have a 50 percent chance and so on down the line until Huntington's disease was either cured or controlled.

No one except another person associated with Huntington's disease can imagine what I went through when I first learned that there was a chance that I, too, could be a Huntington's disease victim. I was married and had a one-year-old daughter. I hated my parents for putting me in that position. I felt my whole life was ruined. My father is now 62 years old and shows no signs of Huntington's disease. So, myself and my four brothers can be fairly certain that we have escaped the disease. But there are SO MANY other people that have not been so lucky. A number of my own cousins have already been diagnosed, or are at risk; Huntington's disease.

Money is desperately needed for Huntington's disease research. Federal funding and support is needed. Research must be done in order to cure Huntington's disease, or even to control it.

Huntington's disease victims require constant medical care. Most times in Federal or State funded institutions. If a cure or control for Huntington's disease was found, millions of dollars would be saved in the long run.

I sincerely hope that the Commission will recommend the money needed for Federal research that is needed to find the cure for Huntington's disease.

THANK YOU!

Denver, Colorado

April 19, 1977

MRS. JOSEPH SNOWDEN
DENVER, COLORADO

APRIL 19, 1977

When my husband began to feel he needed medical attention, he went to our family doctor, Robert Wood, who in turn recommended he see a neurologist, which he did--Donald Ryan, who told him he had Huntington's disease. We felt we wouldn't take just one medical opinion, so selected another neurologist, Stanley Ginsberg, who confirmed Ryan's diagnosis. They both said there was nothing that could be done about it.

We felt we couldn't leave it at that, so my husband entered the V.A. hospital in 1969 in Denver for some tests, and here again it was confirmed he had Huntington's.

There was a young doctor, Charles Johnson, from the Colorado University Medical School, who was in on the case who was very concerned. But here again there was so little that could be done. He was released on the medication Haldol. An appointment was made to see Dr. Arthur Robinson for some genetic counseling and my husband was to report to the Medical Center on regular visits. After the first visit he refused to return--feeling it was hopeless.

He was finally unable to obtain or hold a job as an electrical engineer, having graduated from Colorado University School of Engineering, serving in the Signal Corps in World War II, returning and working for the Bureau of Reclamation for about 10 years, then Sundstrand, Denver Martin Marietta and ECI Consulting Engineers before his health began to fail.

When he couldn't work any more, he became very depressed and went through many stages of mental anguish--and concern of how the family was going to manage financially. I went to work and we were able to get Social Security disability for which we were most grateful. We were able to keep our home and family financially stable for the following six years.

My husband was able to care for himself during that time but this last year he failed mentally and physically rather rapidly. After a bad fall that hospitalized him with a lacerated forehead, I was told he was going to need nursing home care. I was very reluctant to do that until we at least tried

caring for him at home. Having a lot of vacation days saved, I took two days and my two days off. Our youngest son came down from college and covered the weekends and another son was able to cover the other day so we were able to give him constant care--dressing him, spoon feeding him and assisting him to bathroom and walking, etc. This lasted for six months when my vacation days ended and this existence was beginning to tell on the family, so we realized we had to admit him to a nursing home but the financial aspect was frightening. No way could we manage without selling our home or using up what little savings we had.

We had some wonderful assistance from the Social Worker at Lutheran Hospital who gave us suggestions and nursing homes to look into, and ways we might find some financial assistance. Her name is Dorothy Tiedeman. We tried to get him into the V.A. hospital in Denver, but even through our doctor we were unable to. We could not get into the Medicaid program because we had secondary property in a summer cabin in the mountains, even though we had given it to our son to improve and make a home for himself. It was still in my husband's name, so we were desperate, when someone suggested we look into the V.A. and see if we could get financial aid there. We contacted the Veterans Affairs Office of Colorado and Mr. Ralph Ritchie was a tremendous help in this. His concern was heartwarming after so many rebuffs by other agencies. My husband is now receiving a pension plus aid and assistance which allowed us to give him nursing home care and he is now in Garden Manor Nursing Home.

The cost of nursing home care is a frightening thing to the average income family with the possibility of a long stay; without aid, I don't know how they can do it.

The saddest part of all this is the shattering effect it has on the children. When we were married we did not know of the presence of Huntington's in the family. It was thought that my husband's mother had a nervous disease--some said Parkinson's.

We have five beautifully wonderful children whose love and compassion for their father is deep. Two of them are married and two are thinking of marriage, but the disease is putting many obstacles in their paths. One went to seek genetic counseling with his fiancée at the Colorado Medical School and made the acquaintance of Dr. Kimberling doing research on

Huntington's who asked our family to participate in a research program. So when the family all gathered for Christmas this year, we went to the genetic clinic to begin the tests.

It is the only hope they have--research--to lift this black cloud hanging over their heads and give them hope.

This is my prayer.

Thank you for listening.

GAYLE SOERGER
YUMA, ARIZONA

OCTOBER, 1976

My fine and good husband died on December 2, 1975. He was 45 years old; the father of two sons. We had been married 22 years. Though his contribution to the American society was confined to our home town of Yuma, Arizona, he nevertheless contributed far more than most of us. He was no ordinary man, for he touched the lives of many young people through his profession as a teacher and coach. Perhaps his biggest contribution was the way he faced his death; he showed a whole town the kind of strength each of us can have if only we sincerely try. Today we hear much ado about "Death with Dignity." Ed brought dignity to death.

When we first learned of his impending death we acted in diverse ways. Ed would not accept it; I wanted us all to die; our oldest boy figured out that this would possibly come to him; our youngest son chose not to speak. He speaks little even today--eight years later. We moved on to other reactions such as Ed accepting and planning for the future of his family. He checked into his teacher retirement, bought a car for cash (so I would not have payments), and generally purchased necessities and had the house put in order while we still had his income. I went on to college, doing my last two years in 12 months so I could hurry back to my family. Our oldest son eventually turned to marijuana and associations with people who were alien to our way of living.

Denver, Colorado

April 19, 1977

Our youngest turned to alcohol. All of this I kept from my husband since any problem was always magnified in his mind.

Problems like these would have caused scenes and heartbreak beyond what I felt it was worth. I will never know if I handled this rightly when I refrained from sharing these problems with Ed, but I had to make a decision and stand by it. Today the boys seem to be doing much better, but I know they feel the sword of Damocles hanging over their heads.

I feel a great deal of animosity toward doctors for being so ill-informed and challenging me because I dared to question their pronouncements. Some examples: One neurologist stated that women did not inherit HD. My sister-in-law has it. The pathologist who removed brain tissue upon Ed's death asked me when Ed had had the flu which caused HD. Another neurologist asked me why Ed was so unsure of things. A neurosurgeon "experimented" on my husband and made him worse than he had been. I shall stop with that and ask any of you who may read this letter to warn ALL HD patients to avoid the trauma and uselessness of surgery. It can in no way help.

My husband was finally housed in our local nursing home where he received good care from the nursing staff and his neurologist. But, he was being cared for by people who knew him either directly or indirectly. He was loved and admired. But what of those persons who are not loved and admired but have HD? Are they to be treated any less humanely? Complete understanding

MUST be given HD patients and their families. No one who has not lived the nightmare of this disease can ever realize how it feels; therefore, the medical profession has to be totally informed and ready to understand the HD families. If they do not have that compassion, they should stay away from HD. I venture to say they had better seek a different profession altogether.

I believe the government's commitment to fully research neurological and genetic disorders (and all other diseases) should supersede any war and space efforts. I further believe our government should produce a nationally televised program informing our entire country about HD. Great Britain did it last year, and her people know. We are the greatest and richest nation in the world and our people are not informed. I do not want people to go through the humiliation of being labeled a drunk because they have HD or being denied the right to work or have insurance. This is the most insidious form of discrimination.

I feel the sweet memories of a good life with an extraordinary man. But I feel, too, the bitterness of medical ineptness and the terrible agony of knowing that I gave life to two beautiful sons only to wonder which one--or will it be both of them? If I could dare to hope those sons could be saved by research I could be at peace. Help us, in the name of God.

April 19, 1977

LAWRENCE V. STERN, M.D.
HENRY I. YAMAMURA, Ph.D.
PETER C. JOHNSON, M.D.
UNIVERSITY OF ARIZONA
TUCSON, ARIZONA

MARCH 21, 1977

This letter is to express our great interest in the activities of the Commission and to advise you of our willingness to assist in any way that you might deem valuable (including testifying orally, if necessary) in the accomplishment of your important goals. We represent a multidisciplinary team consisting of a neurologist with specific interest in Huntington's Disease (HD) and its consequences (Dr. Lawrence Z. Stern), a neuropathologist (Dr. Peter C. Johnson), and a group of biomedical researchers under the direction of Dr. Henry I. Yamamura, using basic neuropharmacological techniques in an effort to find more effective forms of treatment than are currently available for this disease.

We are presently following over 30 patients with HD at a special clinic which is held at the University of Arizona Health Sciences Center and for which there is no physician's charge currently being made by Dr. Stern. At-risk individuals as well as HD family members are also seen at this clinic to which referrals are made from throughout the State of Arizona. We know of approximately 60 patients in Arizona who carry a firm clinical diagnosis of HD at the present time and are aware of over 300 affected families through our close working relationship with the Arizona Chapter of the Committee to Combat Huntington's Disease.

Our research efforts have largely been based on recent neurochemical findings in HD which have led to new theories and approaches aimed at treating individuals afflicted with this disorder. Clinical pharmacological and neurochemical evidence suggest that activation of synaptic receptors for ACh and GABA may be the most specific and thus possibly the most efficacious therapeutic approach. Acetylcholine precursors such as choline and deanol may have therapeutic efficacy. Development of centrally

active GABA receptor agonists has been hampered in the past due to the lack of specific biochemical or behavioral screening models. The recent development of a biochemical assay for synaptic GABA receptors will enable neurochemists and neuropharmacologists to more rapidly assess the potential usefulness of a large series of compounds as GABA receptor agonists. It has recently been reported that rat striata lesioned with kainic acid are morphologically and biochemically identical to human striata in HD. Most importantly, rats lesioned in this manner exhibit a marked rotatory behavior which may be indicative of an overabundance of dopaminergic input on the lesioned side. Such a behavioral model could also be quite useful in screening for potential GABA-mimetic agents.

With these new biochemical and behavioral techniques and possible prototype compounds it now appears reasonable to believe that we are on the threshold of developing a whole new class of therapeutic agents, GABA agonists. Based upon what is presently known about GABA, this raises the exciting possibility that these agents may be of significant benefit to patients with HD.

Another recent area of our clinical and research work concerns efforts to find a possible predictive or specific diagnostic test for HD. We are also seeking to find a test that might serve as an indicator of likely drug efficacy in HD. Based on the previously reported finding of decreased GABA levels in the cerebrospinal fluid (CSF) in HD patients we have been working with a simple and sensitive assay for CSF GABA which we have used to measure GABA levels in HD patients and control subjects. The radio receptor assay method we are employing involves displacement of ^3H -GABA from the GABA receptors of rat brain synaptic membranes and appears to be specific for GABA in the CSF. Our preliminary results are very encouraging and we are proceeding with more extensive testing of this method which is to be correlated with the clinical course of patients and at-risk family members.

The CCHD has been very helpful in providing us with support for our clinical and research work thusfar. However, it is our firm belief that because of the complex medical, social, and psychological problems encountered by patients with HD and their families it is now necessary to embark upon a much more comprehensive approach to the funding of clinical and multidisciplinary research programs relating to this disorder. This might best be accomplished by identifying active programs already underway and providing them with sufficient support not only to allow them to continue their efforts but to expand them. If there is any way

in which your Commission can help achieve this goal, we feel it would be a tremendous accomplishment - one which would likely accelerate advancement in our understanding and management of this tragic disease.

JAMES R. WAILES
UNIVERSITY OF COLORADO
BOULDER, COLORADO

MARCH 15, 1977

It has recently come to my attention that the Congress has created the Commission for the Control of Huntington's Disease and its consequences and that the Commission has been charged with the responsibility of developing a comprehensive national plan based on the most thorough, complete and accurate data and information available on the disorder.

I feel that the creation of this commission is long overdue, and that information on Huntington's, as well as other genetic diseases in general, needs to be available to the science teaching community in the schools of the country. The educational needs are well-known and a mechanism needs to be set up so that teachers may be informed with accurate factual information. I sincerely hope this is one of the goals of the Commission.

I wish the Commission the highest degree of success in achieving accurate information and dissemination procedures on all genetic diseases.

Denver, Colorado

April 19, 1977

HARRY WARD, M.D.
DENVER, COLORADO

MARCH 31, 1977

Dr. Donald Armstrong, assistant professor in the Department of Neurology here at the University of Colorado, School of Medicine, has acquainted me with the activities of the Commission for the Control of Huntington's Disease and its Consequences. Realizing the need for support for the Commission's activities, I wanted to express my views as Dean of the School of Medicine.

Our experience in this region would certainly emphasize the need for considerable effort in the area of Huntington's Disease and allied neurological diseases. There is an obvious need for improved medical education so that medical students and house-staff might more readily recognize the diagnostic criteria and learn the natural history of the disease. There are obvious needs for education in the area of genetic influences both in research and in dissemination of this knowledge through counseling of affected families. There is a great need for better diagnostic criteria for these diseases. This should encompass diagnostic criteria developed from research in areas of biochemistry, tissue studies and special procedures such as ultrasound, nuclear studies, etc.

There is a great void in the care of these patients whose downhill course is so prolonged and so difficult. This should include the involvement of nursing services and special rehabilitation services as well as educational efforts for the families involved.

We, here, at the University of Colorado, School of Medicine would be most willing to participate in a national effort to step up the biomedical research and education necessary to control this disease. We strongly support your Commission's efforts to develop congressional resources for categorized funding through the National Institutes of Health.

Denver, Colorado

April 19, 1977

MRS. G. F. WARREN
PHOENIX, ARIZONA

JANUARY 10, 1977

I am writing this letter in behalf of my deceased mother, Anna, whom I took to Elgin Mental Hospital in Illinois. At the time she was suffering the shock of having an unknown illness and weaving about on her legs.

She begged me not to leave her more than it took to take tests, which included shock treatments. One lady ran nude while we visitors were there. My mother worked in the kitchen for 15 days, and I rode her the 100 miles to our home. Our local physician advised surgery, for only one kidney was performing. I was advised in his office that she would grow old rapidly as Parkinson's was her disability; but no mention was made of Huntington's chorea or its inherited pitfalls.

I'm 63 and have two sisters with no symptoms, but my brother of 55 has retired from work due to not being able to control his arms. My mother's death certificate does not mention H.D. and none of her relatives (parents, sisters nor brothers) developed it.

I wish there had been counseling to aid my father to assist Mother's feeding and physical needs. He attended her daily and worked at night. I slept with her nightly and worked days.

I regret we were not informed that her mind was functioning regardless of her inability to speak. Her limbs jerked constantly and we did not take her to public restaurants as people stared at her grimaces and trembling condition. We did ride her to see her grandchildren and children and only sister nearby.

My brother is more fortunate in having modern medication to restrain the arm movements. He talks a little, but sleeps many hours due to the medicine. The medicine slows him too much to drive now.

An urgent, quick cure for thiscrippler is a possibility if Congress advances the funds to enable neurological breakthroughs!

Denver, Colorado

April 19, 1977

MRS. G. F. WARREN
PHOENIX, ARIZONA

JANUARY 10, 1977

This letter is typed as a result of my brother Harry's wife and his family's need for advice. He can't type any more so it will be my version of their sorrow.

He is extremely handsome, has been a LCDR in the Naval Air Force and a reserve in that service for the past 20 years after World War II., a pilot. His son is a minister and Harry always has been close to his church. Alcohol and cigarettes were never used.

We noticed his eyes and brows twitching five years ago, but only this year his arms shook; medication to slow them caused him to be drowsy and not alert enough to drive 100 miles round-trip to his employment. So he was forced to retire against his will, as he desired to work until 65 years of age. At 55, his wife drives, keeps him cheered up, and he attends church weekly and visits relatives.

Actually his wife, Margie, is nervous and needs counseling to keep her nerves intact. She never thought of sending him away, but encourages him to keep a little active. He has started to read again. The medication for slowing the arm movements keeps them very quiet, and there is no visible sign of Huntington's, except for the twitch of face and eye frequently. At the time our mother had H.D., her limbs constantly moved, as no medicine was available. She lost her speech, but Harry forces himself to speak out, and at times it irritates his wife with that problem of slowness.

I thank CCHD for making us aware of the swallowing condition, and that his mind is functioning in other respects. It hurts me so each time I see him and know I can do nothing to destroy his killer. His doctors never knew, or did not tell him, his malady. CCHD brought H.D. to our attention, as we believed our mother had Parkinson's. Harry's two children and four grandchildren may get help soon; but we desire his cure immediately if Congress will sponsor scientists in this field and bring a halt to all this unhappiness in our U.S.A.

Denver, Colorado

April 19, 1977

ROSALIE WIESEN
PHOENIX, ARIZONA

SEPTEMBER 21, 1976

My mother has been afflicted with Huntington's disease since I can remember. As a child and teenager, I saw my mother's frustration and anger with her increasing loss of muscle coordination, and her depression and occasional outbursts of violent anger. No one in my family knew what was wrong with my mother, although we saw two of her sisters developing similar symptoms and I knew my grandmother had the same type of "nervousness." My mother's condition was never discussed, and as a child I had the feeling that the truth would be too terrible to deal with.

Now my parents are separated from their children, who have all moved to different areas of the country, and my father cares for my mother at home. Their life is extremely grim and loneliness is an ever present problem for them. They are in their 70's, so their problems are compounded by the usual problems of the aging. My mother is in the advanced stage of Huntington's disease--and lives in a nightmare of suffering and hopelessness. She can barely walk, and falls down frequently. Everyday activities require enormous effort--eating, talking, dressing, washing. My father refuses to do for her what she can do herself, but he complains that she needs constant prodding because of the apathy and weakness which the disease causes. He refuses to get outside help to care for my mother, because he has seen my aunt become totally helpless after years of nursing care at home.

When I first learned about Huntington's disease I was 25, and learned that I have a 50% risk of developing the disease, I found it difficult to accept, and it has not gotten any easier as I have seen my mother's deterioration. The knowledge that I might have the H.D. gene certainly influenced my decision not to have children while I was married. While I would like to remarry eventually, it worries me that my future is so uncertain. It is not something that I dwell upon, but it is a very persistent fly in my ointment.

Denver, Colorado

April 19, 1977

LINDA WILLIAMS
DENVER, COLORADO

APRIL 8, 1977

My name is Linda. I am 23; my husband, Michael, is 25. We have two little girls ages 3 and 4. We recently had it brought to our attention that Michael might be a potential H.D. victim.

We have just about come to the end of the road. We have tried everything.

All we have been able to gather as evidence that it may be in his genes is the word of a self-confessed perpetual liar, his mother. She never even had it verified that what she had was H.D. before she died. We have not been able to track her bloodline back to her mother, who supposedly died of H.D. She died at the age of 67 and only a few years before, she was seen in a hospital but did not have any of the symptoms of H.D. So that leaves us nowhere. We've tried everything in our power to get more medical information on Michael's grandmother. We have been told that we will have to get a family doctor in Michigan to send her medical records to a family doctor in Denver. Well, in the first place we don't have a family doctor in Detroit; in the second place we don't even know what hospital she died in.

I've worn my fingers to the bone writing to every hospital that I can get the address for in Detroit and so far no one has been able to help me.

My first concern is to find out one way or the other whether H.D. is in my husband's bloodline. My second concern is to protect my children and their children from H.D. I would like to have more children but at this point I am unsure of what my children might have in their future. At this point my only hope is that research may find an answer.

I pray that in the near future there will be a cure for H.D.

ANONYMOUS
RIDGWAY, COLORADO

MARCH 21, 1977

I have taken time off from work to concentrate on the impossible task of this letter. . . .a subject. . . .a condition and constant fear--HD. It cannot be compared even to a snake pit for the feeling hauntingly strikes always by day--never misses a night. My life as an at-risk person (female--age 41) has not been a life, rather an act, an act of pretense, exclusion, a bitter reality, an act in which anything remains easier than the truth.

What does an "at-risk" person "live with". . . let me delineate just a portion of the facts:

1. Fatigue--an overwhelming, consuming fatigue. I need all my energy to be what I am today.
2. Career--as a special education teacher, my job has always been jeopardized by the truth.
3. Health--Life Insurances--show me a policy that would not be canceled if the truth were known?
4. I am a twin--so the horror is doubled....which one should be sick?
5. Divorce--the HD factor was present in the divorce of my parents, and the divorce of my twin.
6. Relatives--I have adolescent nieces and nephews who have not been told and constitute a conflict of the truth vs. silence.
7. Marriage--I am single as a witness to a broken engagement when the truth was spoken. I remained unmarried now as I do not believe this burden can be successfully shared.
8. Adoption--turned down as an unsuitable candidate with a history of "at risk."
9. Fear--a stockpile of pills if suicide becomes the only answer to symptoms.
10. Participation in HD Chapters--Anonymous.

11. Participation in research--I have done this but not without the turmoil of wanting to contribute, but at the risk of non-confidentiality.
12. Coping Mechanisms--outbursts of anger followed by remorse. Guilt at imposing the burdens of my adversity on the few close friends with whom I am fortunate enough to be able to share this intolerance. Drinking--to kill the pain of fear.

There must be National Funding for medical research to determine etiology, early detection, control of symptoms, and ultimate cure. Funding through legislation to expand educational understanding of all appalling ramifications of HD. Funding to make comprehensive health care, nursing homes, and insurances available when this tragedy occurs. Funding is needed to expand the role of genetic clinics so that this terrible strain will not continue through procreation.

In summary, I implore you to "ask someone unburdened by fate to contemplate the greater burdens fate has imposed on another." This Commission for the Control of Huntington's Disease and Its Consequences holds the greatest potential--that of hope. I might try again to find a life of my own--as opposed to this "existence" of many rooms and many doors that are not presently mine to open. "I have learned that success is to be measured not so much by the position one has reached as by the obstacles which have been overcome while trying to succeed." Booker T. Washington.

Please help us!

Denver, Colorado

April 19, 1977

ANONYMOUS
DENVER, COLORADO

MARCH 31, 1977

I am 81 years old, second from the oldest in a family where seven grew to adulthood. Of this family five have contracted Huntington's disease. Three of this number could have passed on this disease.

The history of my family was told to me by my mother, and may be of interest to others.

My paternal great-grandfather came from Wales. It was thought there was some stigma attached to him, as each month he was sent a check from his family in Wales sufficient to his support. He married and had two sons. He was considered quite a sportsman and in his early 40's was killed in a sulky race, leaving his widow and sons. His widow placed her sons in foster homes and returned to her parents. One of these boys, who was my grandfather, was taken by a family of the name of Spencer. He married and had four sons and one daughter. It is known that my grandfather had all the symptoms of H.D. but may not have known the name of his disease. Of this family of five, three sons had H.D.

My father married and had seven children who grew to adulthood, three having died in infancy, and hopefully there were no illegitimate children to carry on the curse of H.D.

My oldest brother, eight years older than I, was the first to have H.D. He had one daughter who did not inherit H.D. My brother died in a sanitarium because of choking on a piece of bread.

A brother two and one-half years younger than I began showing signs of H.D. in his 40's, but lived a comparatively normal life, serving in the armed services in two wars. He died as a result of an accident in a V.A. Hospital in Arizona.

Next a sister began showing signs of H.D. when in her early 30's. Because there wasn't anyone to care for her she was committed to a state mental hospital where she died at the age of 50. She did not belong there and spent her time trying to escape so that she could tell the authorities the condition of this institution. When she died at the age of 50 the autopsy showed her to have the body of a 90-year-old woman.

A younger brother married and had four children. From seeing the other members of the family with H.D. he had such a fear of inheriting it he committed suicide. There are at least 13 grandchildren who could inherit H.D. There were no tests taken to confirm or deny this and if none of the four do inherit H.D. then one can assume that this brother died because of the fear rather than the disease.

The youngest sister is still alive but in the last stages of H.D., and she is cared for in a nursing hospital. She married and has three children. Her husband has assumed the financial care of her but now in his early 60's is breaking under the strain, and he writes us that he plans on getting a divorce and going away to try to regain his health and start a new life. My sister shows few signs of reality. The choreic movements have largely been controlled by drugs but she has no use of her muscles, cannot talk, and cannot even feed herself.

No members of a family where there is H.D. can be free of it, of the anguish and heartache. It was my dubious privilege (?) to commit my father, one sister and a brother to mental institutions. They did not belong there but there was nothing else to do. They were not insane but were forced to live their lives with those who were.

God bless Marjorie Guthrie for starting a movement to control this fast mushrooming and insidious disease.

ANONYMOUS
AURORA, COLORADO

MARCH 21, 1977

My mother first evidenced symptoms of Huntington's disease at about age 43. Her physical symptoms were a constant wrinkling and twitching of her brows. Her first sign of mental deterioration was a lack of concern for the family and its needs. Our family home burned in July, 1934, and the new house was ready in December. She was interested in "fixing it up" but not as she would have been if normal. She became more and more unreasonable as time went on and insisted on visiting and caring for her terminally ill sister in California in 1937.

Our mother was an accomplished pianist, an inveterate reader and socially active. She was also very active in church work and was the president of the local school board for 17 years.

As her faculties deteriorated she lost interest in the piano and in church although she still enjoyed going out. In fact, one of the manifestations of Huntington's seemed to be a very restless condition--she could not be satisfied with any one situation for any length of time. This resulted in her insisting that she and our father "commute" from California (where we lived) to Colorado and back several times before we would not let her return to Colorado with her husband for his own safety. She was increasingly violent toward him. He was a victim of emphysema and unable to cope with her.

We, her daughters, then cared for her in our homes in California until it was necessary to put her in a nursing home. She was not satisfied there, of course, and ran away several times--she also created scenes which could not be tolerated. It was finally necessary for us to commit her to a state hospital in 1947 where she remained until she died in 1948 at the age of 56 as a result of peritonitis.

Our long and frustrating search for a correct diagnosis of her condition led us to many doctors, and interestingly, our family doctor in Longmont really came closest to the cause when he determined she had St. Vitus Dance, a form of chorea, with mental deterioration. We finally took her to a neurologist in San Jose, California, who told us she had Huntington's

chorea as it was called at that time. His diagnosis was confirmed at Sanford-Lane Medical School in San Francisco. She spent ten days there, taking extensive tests. We could tell them, by way of family background, that her father evidenced the same symptoms before he passed away in 1929 (our mother recognized the fact that she was like her father). His mother, our great-grandmother, died in the state hospital at Pueblo, Colorado.

It was our experience that even some of our mother's closest friends believed her to have a drinking problem and that was also the case in our grandfather's situation, although both of them were tee-totalers. Her family (sisters) would not accept the fact that this was an inherited condition. When the youngest sister, who did drink, started showing symptoms the family attributed them to alcoholism. Many years later a doctor whom they trusted confirmed the H.D. diagnosis.

Of six daughters born to our Grandfather and Grandmother, two developed H.D. Our aunt is still living in Longmont and is cared for by her husband. She is nearing 75 years of age. Two of the other daughters died at relatively young ages, so we do not know whether or not they would have developed the disease. None of their children have evidenced any symptoms of H.D. so far as we know. We have one brother who does not show any signs of it.

In our family, H.D. seems to have passed from mother to son to daughters. We have not researched our grandfather's family but this is one of the things we plan to do in the near future so that we will know something of its background.

We consulted a geneticist at the University of California, Berkeley, to find out more about the disease. It was there that we learned about the 50-50 chance of one or both of us contracting it. We had married and had children so at this time the period of waiting and apprehension began. One of us was in her middle thirties, the other in the early forties. Since we are now in our fifties and sixties with no manifestations we can begin to believe that we are "home free."

Denver, Colorado

April 19, 1977

ANONYMOUS
PHOENIX, ARIZONA

APRIL 18, 1977

I am writing this letter in hopes of gaining support for Huntington's patients and their families. My father died of the disease. He was from a family of six children. Three died of Huntington's disease and the other three have led normal lives, showing no signs of the disease. All three Huntington's victims had children before they showed signs of having the disease. One of these offspring got the disease at the age of 35. There are still four more offspring at risk. My father got the disease at the age of 40 and his father at the same age before him.

When my father started showing signs of the disease, he was thought to be an alcoholic, insane, etc. My mother and father refused to talk about the disorder. Mother would never admit to friends, relatives or her children that my father had Huntington's and that his two sons stood a 50/50 chance to have the disease.

My brother is now 52 and I am 50. To date we have neither one shown signs of the dread disease.

It is our sincere hope that help will be forthcoming to the families of Huntington's victims on a national basis from Congress. They have a great burden to bear. And, of course, all of us hope and pray that help for the Huntington's victim as well as a cure is coming in the near future.

Please help us all.

ANONYMOUS
ARIZONA

APRIL 15, 1977

My beloved husband died of Huntington's disease at the age of 39 years. We were married in 1919, when I was 17 and he was 18. He was a handsome youth. We lived with his parents at the beginning of our marriage, and his mother was afflicted with the disease at that time. She died in 1927. His grandmother and an aunt died from Huntington's, and an uncle committed suicide when he learned he had the disease.

At this time we did not even know what the ailment was called and no treatment was given. The unfortunate victims were hidden most of the time. It was considered a disgrace to mention their problem. Many times they were mistaken for drunk and arrested if alone on the street.

My handsome husband and I had a happy life for four years. During that time we were blessed with a son, a beautiful baby who died at the age of three. Then we were advised by a doctor to have no more children. This made the baby's death more tragic than ever. My husband never fully recovered from the shock of the baby's death. Shortly after this, he showed signs of nervousness, and I've often wondered if this bereavement hastened the progress of the disease.

He would never admit he was afflicted, although the disease progressed rapidly. He lost his good looks, could not hold a job, and was accused of drunkenness many times. So he took to drink. He was very popular in his youth, admired by both men and women, and a friend to all; but they deserted him when he needed them the most.

Finally he was placed in a mental institution. When the orderlies came to take him away, he resisted with all his feeble strength and cried out for me to help him. I will never forget this dreadful event. I tried to take care of him until he became dangerous to himself and others before I made this dreadful decision; and to this day, I regret it. He refused to eat, and beer was all he wanted. He died of starvation after 18 months in the mental institution. God bless his soul.

His sister was not affected by this inherited disease. She was 62 when she died.

I am thankful that these unfortunate people can obtain help now. I hope the good doctors and science will soon find a cure for Huntington's disease.

Others in my family have requested that our name not be published, but thank you for this opportunity to tell you of the effect of Huntington's disease on our lives.

Denver, Colorado

April 19, 1977

ANONYMOUS
TEMPE, ARIZONA

APRIL 15, 1977

I am 29 years old and my mother was diagnosed with H.D. when I was 16 years old. I have two younger brothers and one younger sister. I am fortunate in that at least I am able to remember my mother as a normal, happy, healthy human being. Unfortunately, my brothers and sister can't.

As with so many other cases, my mother's mother was not diagnosed as having H.D. So, when my parents got the news of my mother's disease, it dealt a terrible blow to our family. My father turned to alcohol and drugs and is still the same today. My two brothers ran away from home and one is now a heavy drinker.

I am married and have two children. Since I am approaching the "crucial years" (35-40) I am scared. Scared to death! I pray each day that maybe I didn't inherit the deadly gene. I honestly don't know what I would do if I did get it. I carry horrible memories of my family and what life was like after my mother's diagnosis. I swear that will not happen to us. I don't know how I will rectify it but I will. My children will not remember me as a twisted, distorted shell that once used to be their mother.

I also pray each day for at least a control drug, such as insulin for diabetics. However, sometimes I feel that it is an exercise in futility. Immediately after my mother's diagnosis she said to me that she knew there was no hope for her but prayed for hope for me. I don't ever want to repeat those words to my daughter.

Los Angeles, California

April 15, 1977

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P R O C E E D I N G S

M. WEXLER: I want to welcome all of you to this hearing for the Commission for the Control of Huntington's Disease. As you probably all know, the basic charge of the Commission is to make a comprehensive study of the state of the art of medical and social management of Huntington's disease in the United States. We're going to investigate as well as we can the problem of Huntington's disease, its consequences in terms of the social and economic impact on families and on government. We're going to try to develop a comprehensive national plan (supported by the United States Government) to solve the problem of Huntington's disease. Hopefully, all of you will be able to give us not only a statement as to the kinds of problems that you face if your family is involved, but those who are uninvolved can give us suggestions, advice, information that will then turn out to be usable in the kinds of recommendations that the Commission ultimately will make to Congress and to the President.

Before we begin the actual taking of testimony, I want to introduce the people who are up here on the platform. I have on my right Mrs. Marjorie Guthrie, who most of you know. Marjorie has been perhaps the prime force in the United States in the development of a program to solve the problem of Huntington's disease. She's the Chairman of this federally established Commission, and she is the head of the Committee to Combat Huntington's Disease. She is a prime mover, and I think most of you know that. Those of you who have not yet met Marjorie might in the course of this day make it your business to say hello to her. I think you'll find her both charming and informative.

I always forget to introduce myself. That's on the assumption that everybody knows me, of course [laughter]. I'm Dr. Milton Wexler, and I'm the Vice Chairman of this Commission. I'm going to chair the meeting today in the taking of the testimony. Marjorie and I kind of divide up the chores of the Commission, in terms of Marjorie being very deeply involved in the problems of the social management for patients who are involved in the disease and my interest over the years being the development of biomedical research to try to find a solution at a scientific level for the management and the cure of this disease.

On my left is a person who I know all of you know without any question, but I'll introduce her anyway, Jennifer Jones Simon, who has been extraordinarily kind and extraordinarily interested in relation to the problem of Huntington's disease. She's been a long-time supporter in this area, and in general, over a great many years (even before the problem of Huntington's disease came into focus in this country), has been deeply involved in the problems of mental health and health in general. I must say that the Commission is extraordinarily proud and pleased to have her as a commissioner.

To Jennifer's left is someone who is extraordinarily vital to the Commission, and particularly vital to me--she happens to be my daughter--Dr. Nancy Wexler. She is the Executive Director of the Commission. I always like to think of it as a kind of reversal of roles because now I can very proudly say that Nancy is my boss [laughter]. She is in title and (although she doesn't think so) in fact my boss and tells me what to do as Executive Director and as daughter.

Let me say a word about the procedure for the hearings. We're going to run straight through until 5:30, at which time I understand we're going to have a break for dinner, and we'll resume at 6:30. The witnesses scheduled to testify are going to be called in groups of three or four, and they will take their positions at that table, which will be the witness table. I'd like to request that as you testify you state your name and address. If there is some kind of organizational affiliation, please give that organizational affiliation prior to commencing your statement.

I'd also like you to please note that there are two microphones, a tall one and a short one, and they serve different functions. The tall microphone is the broadcast microphone for this room. The short one is for the purpose of making a recording of the testimony because, as you probably already know, the testimony will be transcribed and all of your words of wisdom and advice will then be forwarded to Congress as part of the whole document that we are preparing and will be some of the evidence on which Congress will ultimately act in helping to support research and care for Huntington's. I hope that you'll be able to speak directly into the tall microphone at least, but hopefully into both, so that we have not only everyone hearing you but every word of your statement for the record. I think those are all the instructions that I have.

We have a kind of schedule for the testimony. We may have to interrupt the precise scheduling that we have at times for certain of the scientists and doctors who have consented to come here and may be under some pressure to get back to patients. We're going to try to keep the schedule that has already been established. But if we mess up in a few minutes, you'll understand that we're trying to accommodate some doctors who are urgently busy.

Have I omitted anything? Is there anybody up here who wants to add a statement before we begin?

GUTHRIE: Milton, I'd just like to say a word. I just want to add, since I'm on the social management side and my concern is with people, I think that some people are frightened when they think of coming forward and speaking into a microphone in this formal setting. I want to beg you, would you pretend that you walked into your Congressman's office and you sat close by, and he looked at you and said, "What's your problem, and what can I do about it?" and try to put yourself in that position, as if you're talking one to one. I think you'll find it a lot easier than trying to make a speech to everybody.

M. WEXLER: I only want to make a slight amendment. I think, at least, if Congressmen scare you--just like you came up and talked to me. Okay?

GUTHRIE: That's even better, yes.

M. WEXLER: Or you're in your own living room talking to your kids, or whatever. You don't have to be terribly eloquent, or grandiose, or anything. I want you to know that there are an awful lot of people who are tremendously interested in your problem, and they're terribly eager to hear what you have to say. Whether it's organized, disorganized, eloquent or not really doesn't matter. Somewhere along the line, if it isn't clear in any way, believe me, everybody up here on this panel will try to help you. We'll ask questions, and we'll try to clarify it.

If there is no further comment that anybody wants to make here, then I would like to call on Dr. John Menkes, who is Clinical Professor of Psychiatry, Neurology, and Pediatrics at the UCLA School of Medicine. Dr. Menkes.

TESTIMONY OF
JOHN MENKES, M.D.
CLINICAL PROFESSOR OF PSYCHIATRY,
NEUROLOGY, AND PEDIATRICS
UCLA SCHOOL OF MEDICINE

MENKES: Ladies and gentlemen, I don't think I need to tell any one of you what Huntington's disease is and what its manifestations are. Perhaps, however, I should stress the importance of the disease beyond you who are gathered in this room. As you know, Huntington's disease is a disease that is transmitted hereditarily (that is, from one generation to the next) and is a disease that manifests itself by a progressive loss of brain function. As such, it is a very common disease by itself. There are many families in this area and in other areas where there are as many as 50 people who are threatened by this disease, others where there are less, some where the number of people threatened by this disease is not known. This, by itself, renders this a condition of importance.

However, I should stress that Huntington's disease is also a prototype (an example, so to speak) for a number of other illnesses that manifest themselves similarly. For instance, there are a number of illnesses in which a loss of brain function appears late in life--they're called the dementias, Alzheimer's disease, the various forms of senile dementias--where the clinical picture (that is, the picture that the patient manifests to us who are doctors) and the pathological picture (that is, the picture that the brain shows when it is examined under the microscope) are similar to that of Huntington's disease.

Thus, in these conditions we hope that once we understand Huntington's disease and can control it, we can then readily progress to the other diseases that are somewhat similar and do the same for them. I mentioned the dementias. I feel that some of the schizophrenias that one sees, in which there is also a progressive loss of mental functions, are similar in their manifestations to Huntington's disease. As such, then, Huntington's disease has a considerable importance beyond the numbers of patients and their families that are directly affected by it.

Having this opportunity to talk to the Commissioners here and to you here, what can we say? Where we are and what we need to

finish, so to speak, a job that we started many years ago. We are, I believe, on the threshold of understanding the basic nature of this illness. We are at the point where we can see that this is, in a way, quite unlike any other illness of the brain that we have yet understood. We are almost at the threshold of being able to detect the disease before the neurological symptoms appear (that is, before the brain begins to deteriorate). We are, therefore, perhaps also at the threshold of being able to prevent the disease by being able to tell in an unborn child whether or not he or she will ultimately be affected by the disease. We are also, I believe, on the threshold of being able to conceive a mode of therapy for the disease once its first manifestations (that is, its first ravages) have begun.

What do we need to continue this job and to finish it in the way I believe polio was finished? Polio is a rarity. I myself remember very distinctly back in 1952, when I started my pediatric training, the devastation and the frightening aspects of polio. No one now, unless they're older than I am, will remember what polio meant in those days. Polio, as a disease, is finished. Tay-Sachs disease, which is a genetic disorder, is almost finished as well. Even though we do not have a cure for the disease, we can now isolate those people who carry the gene, instruct them, and also ultimately prevent the birth of individuals who will become affected by this disease. Therefore, I believe that Tay-Sachs disease will no longer be of any significance a few years from now, and it is merely a matter of public health measures to prevent this disease from continuing.

What do we need to put Huntington's disease in this same category? First of all, it's always easiest to say we need money. That's very easy. Everyone will come before Congress, be he a logger or be he a scientist, and want money. I think we should be more specific than saying that we want money. I believe that we need to continue to instruct young scientists about this disease and make it interesting and attractive and reasonable for them to continue their career in trying to study and combat this disease. This means supporting them through their younger years, through the years when it is difficult for them to get support, providing them a means of livelihood--after all, one cannot live by ideals alone, at least not adequately--and allow them to spend a given number of years starting out their research on this disease. We

need support for young scientists and instruction for young scientists and get them involved in this area.

We also need support for the established scientists that are working on this. Of course, these people, like myself, are able to write applications to the Federal Government and other private institutions to get money, support. It is getting harder and harder, of course. We have to compete with those people who are writing equally good grant applications for support in diabetes, cancer, and stroke. The thing that we ask here, I think, is that we should be treated equal to them, if not perhaps, hopefully, a little bit better than the other areas.

Finally, I think that we need to consider the possibility of setting up some research center, be it in Washington or be it elsewhere, that is devoted to the study of Huntington's disease, to an integration of all the disciplines that are bearing upon this disease and are able, by their knowledge and their background, to help us solve this disease.

I should also add parenthetically that it would be very nice if the Federal Government would allow people who have established themselves and who are working on this disease the freedom to concentrate on the scientific work so they would not have to (as myself) make a living by seeing patients. This, of course, detracts from the intensity of intellectual effort that we can devote to this problem and also dissipates our energy.

Hopefully, if all this is done, or if only a part of this is done, we will be able to progress as much in the next seven years as we did in the last seven years, when in 1970 I first met Marge Guthrie and she got me involved in working on this purpose. Thank you.

M. WEXLER: Does anybody up here want to ask Dr. Menkes any questions?

N. WEXLER: Could I ask just one specific question? How would the granting system need to be altered so that you could focus your work full time on research? Is it the amount of the grant you would increase, or structure the grant differently?

MENKES: No, it would be what used to be present. There was career investigator support. This was at one time a federal slot for career investigators where, let us say, three- or five-year salary support was given to a university for a career investigator to work full time on a project.

N. WEXLER: That exists no longer?

MENKES: I do not believe it exists any longer.

M. WEXLER: In connection with the recruitment of the young scientists, would you have anything to say about the value of a workshop program?

MENKES: I feel that the workshop program is what brings Huntington's disease before the young scientist, before the young biochemist, before the young immunologist who has the basic training that is important in Huntington's disease; but who, because of the training being in the basic sciences, has never even heard of the disease. I think this is how he can be attracted to this area.

M. WEXLER: I wonder if you'd say anything about the value, if you think it has a value, of any kind of specialized clinical research centers? Do you think that would be applicable here?

MENKES: Clinical research. At this point, I am not sure how much more we can offer. The direct approach in a patient is so difficult when the disease is located inside the brain. Also, our attitude towards clinical research has changed considerably, and it is becoming harder and harder to really perform clinical research. So unless and until the atmosphere and the restrictions that have been placed on clinical research are altered, I don't believe there will be much hope in that direction.

M. WEXLER: Any other questions?

N. WEXLER: Would a registry be of any use to you in your research or in research that you know of, a registry of HD patients?

MENKES: I think it's of very important use. I think that it's a necessity.

N. WEXLER: How so?

MENKES: I think, first of all, in contacting all the patients and providing enough patient material to do the studies which do require a patient. We can't always approach the same patients and say, "Please give us a piece of skin." They won't have any skin left after a while.

N. WEXLER: I'm glad there's so much research.

M. WEXLER: John, thank you very much. We appreciate your coming and talking with us. [Applause.] I think we're going to have to have a program in which the applause will come at 10 o'clock; otherwise we're going to have to applaud everybody who steps up here.

I'd like again to make some deviation in the planned program and call on Dr. Eugene Roberts. Dr. David Barklay, I think, should come up. David, do you want to come up, too?

N. WEXLER: Since we are altering the schedule, if there are certain of you that have particular scheduling problems or times when you have to leave to catch planes or go to work or whatever, would you see Mr. Booth. He's over in the corner with his hand raised. Thank you.

M. WEXLER: Since I'm not sure that either or both of you were here, when you begin your testimony would you please give your full name and affiliation so that we have that for the record. The taller microphone is for this room, and the shorter one is for the taping. If you would put the two together when you're talking, it would be very helpful. Why don't we start with Dr. Roberts.

TESTIMONY OF
EUGENE ROBERTS, M.D.
DIRECTOR
DIVISION OF NEUROSCIENCES
CITY OF HOPE NATIONAL MEDICAL CENTER

ROBERTS: I'm Eugene Roberts, Director of the Division of Neurosciences at the City of Hope National Medical Center. Our division is devoted entirely to work in basic neurosciences. Insofar as patients in my particular division are concerned, I think the highest level of patient is the cat. We are working very

closely with Dr. David Comings, who you will hear later, who is the head of our Department of Human Genetics and works directly with patients with Huntington's disease and also participates in fundamental laboratory work related to that. Our work is dependent on obtaining material, advice, and guidance from him.

I'd say that Milton and Nancy Wexler are absolutely and directly responsible for my even knowing what Huntington's disease is and participating in the research, and I'm deeply involved now. Their catalytic function in getting research going has just been absolutely phenomenal. I think that they deserve a fantastic amount of credit, and I think that it shouldn't end here because the techniques they've used and the way they've elicited interest from many people is a pattern which could be repeated over and over again in the study of many important problems involving human beings. As far as I'm concerned they are A-number one and should receive a fantastic amount of credit for whatever has been achieved today.

With regard to my own interests, the fundamental cell properties of membranes and growing cells, I, together with Dr. Barklay and Dr. Menkes (who spoke to you just now), did come upon some properties of these cells grown from the skin of patients with Huntington's disease and normal individuals (comparing them) which looked to us as though they're very important leads in studying what might be wrong with these cells. That work hasn't progressed to the point where we know exactly what molecules are involved, what the diseased molecules are and how these could be manipulated exactly for diagnosis or for treatment. At this point there's a lot of hope, though. So we (and others, hopefully) can train the armamentarium that's available to people who use chemical, biological, physical techniques in studying these problems. I must say that these techniques are very advanced now and give us great opportunities to investigate further, to get some kind of issue at that level.

I'm very hopeful, but I'm not going to guarantee when it will occur, because usually that kind of discovery at that level occurs in the head and with the hands of one individual. If I knew when to predict and how to predict where it would be, I would be doing it. We're very hopeful, though, that these extremely brilliant young people who have been brought into the program and interested (who

before, as Dr. Menkes said, didn't know what Huntington's disease was), that among them--or maybe in our own laboratories, we don't know--will be an individual who will penetrate to the real, fundamental molecular defect. We don't even know what the consequences of that knowledge will be, so I must plunk for basic research at that level, in the laboratory, with the individual often working alone with his thoughts and taking chances. But that's just one aspect. I think that we have to work both ends toward the middle.

The other end is something I'd like to share with you that I'm very excited about. Other people, and even Dr. Barklay, I think, may not be as enthusiastic as I am about this. But being an enthusiast I have to share my enthusiasm, and sometimes I'm right and sometimes I'm wrong. I think I've been right about 60 percent of the time--I mean, about enthusiasm [laughter]. Anyway, there are some techniques worked out now--and we heard a report about the beginnings of that at our last meeting--in which it is possible to really see, without any danger to the person, some of the fundamental processes that are occurring in the brains of these people. It has kind of a terribly complex name. It's called Positron Emission Computerized Tomography. What it involves is injecting very small amounts of some substances that give off a harmless form of radiation which can be detected, and by using other techniques that were developed for looking at essentially section after section of brain, it is possible to see now what the metabolism is with regard to glucose in very discrete parts of the brain, what the circulation is there, and also some aspects of protein metabolism. It really blows your mind. It's really just one of these space-age discoveries. We are in contact with these individuals, hopefully that it will be possible to work with them.

I can foresee that it really would be possible to follow an individual patient's progress, or whatever it is, the effects of the treatment of various drugs, or whatever is happening to them, in this way without any danger to them. This whole bit of equipment costs about a million dollars to install in a place; and, of course, you need highly skilled individuals. I can't help but feel that that kind of work should be pushed immediately.

Let's come to the third part, what I call the R & D phase. This particular thing turns me on because I think we can begin to see inside a person's brain without doing anything to them that's

harmful, or obnoxious, or that will have any effect on them at all except spending some time lying on their backs in the laboratory. There are a number of possibilities (in drug treatment and even in things like some of the vitamins that are available) to explore. I think that's sort of R & D, research and development things. There are some drugs that are known to help some patients with Huntington's disease; for example, haloperidol. There are some drugs which affect the cholinergic, or acetylcholine, system that have been claimed by some people to be helpful. It's possible, as far as I'm concerned, that maybe relatively large amounts, say, of vitamin C and vitamin E might be helpful. If not, they certainly wouldn't be harmful, and I think it would be something to test, and so on.

These things are relatively mundane. I mean, they don't turn me on, as a person who likes to work with the molecules in the laboratory. I think they're very important to pursue. There are people who are competent in this field and who are turned on by these kinds of things. I think their interest should be elicited, and funding should be available.

The last thing I want to discuss with you is this: that now given these things (basic research, research and development, and direct application of some techniques--and I just mentioned things in sort of a very impressionistic way), what do you do now if you think you have a lead, or if there is a consensus among a number of people that there is a lead. To me, that's one of the world's most frustrating things, to just sit around and then apply for a grant, or hope that somebody somewhere is going to say, "Well, I, too, am interested, and maybe I'll work on it if I get a grant," and this goes on a year and a half. You know, it doesn't take many years, or year-and-a-half periods, to use up a person's scientific lifetime. I think there should be some mechanism when there is a consensus--and I don't mean in one person's mind, I mean a relative consensus, "Yeah, we got a lead that should be explored and several people are interested"--that you should be able to push forward with full speed without all the things that kill us.

I have quite a bit of gray hair, and I'm sure a good share of it is due to the worry and concern about writing grant applications to keep my associates and myself going in the laboratory. I work in a very fine place where my full salary is paid by the institution, mind you. A lot of people are even having to pay a good portion of

their salary out of the grants in our major universities, including Harvard and hallowed places like that. I think it's ridiculous to be in that state of affairs now when the total amount of money that would be required, say, for this kind of push is just a fraction of one of the bombers that's being legislated about and going to be constructed. I don't know how to change this thing around. I think it's one of the world's most frustrating things. I try to think of it as little as possible because it's enough to just drive you crazy when you think of what could be done with so little and how little is being done with so much in some other places.

Essentially, I'd just like you to know where I stand and where I'm at with regard to my feelings and this field. I'll be glad to answer any questions.

GUTHRIE: Dr. Roberts, do you have this machine that you need for tomography?

ROBERTS: We don't, but it's at UCLA, and we've heard that it's going to be operational shortly. They are very interested in Huntington's disease. You see, as far as their setup now, we were told that with all the things they have to do with this machine they are going to be able to run about eight patients a day altogether. As you can imagine, Huntington's disease is competing with stroke, it's competing with cancer, it's competing with all other things for this kind of facility.

GUTHRIE: Right. That was my next question, though. Is it being used then on other disorders?

ROBERTS: Oh, yes. This is a technique that will tell you the things that are going on in the liver and in the kidney and in the head. Of course, a lot of things go on in the head. They're going to be doing a very important project in schizophrenia because in some types there is no question that there's something physiologically wrong in the circulation, and people will be looking at it.

After some preliminary work, which we hope can be arranged without a big input, if we identify this as a useful approach for Huntington's disease, then there should be such a facility just for Huntington's disease. I'm sure that for some time to come the Huntington's problem could occupy such a small--it isn't a big

facility in terms of size, but it is in terms of the armamentarium and the type of people required.

GUTHRIE: I think what I'm trying to get at is, would you like our Commission to recommend that we either add additional tomographic--what do you call it?

ROBERTS: That kind of equipment.

GUTHRIE: Yes, equipment.

ROBERTS: What I'd like to do is to defer that to the people who are really doing it, and for you to get that answer directly from them. For example, Dr. Louis Sokoloff (at the National Institute of Mental Health), Dr. [Cool] and Dr. Phelps (at UCLA) could talk to you. The last time I talked to Dr. [Cool]--he called me about a week ago--he was very enthusiastic about the potentiality because the particular region that is suffering in Huntington's disease is very well visualized by their technique. So, you make a halting start. It's nothing I have anything to do with. This is not my technology. I don't even understand the mathematics involved. But the potentialities of it just floored me when I recognized what was going to be available. I think the experts should answer that question. My feeling is that if it is judged that it's worthwhile, then I would certainly plunk for having these people have another setup in their hands, in their place, with their guidance, which could be devoted to the study of Huntington's for a time.

M. WEXLER: Gene, I got the impression that you were trying to suggest another mechanism aside from the grant mechanism. Were you referring to a contract mechanism?

ROBERTS: I was referring to a contract mechanism, but different from the contract mechanism as I understand it to be that the NIH has now. Let's put it this way, a kind of mini-Manhattan Project, where a good, honest, impartial, and hard-driving directorate is appointed and then given a sum of money to do the job, and not a contract where you say, "Yeah, I want to write a contract proposal and then give a three-month report" (every three months spend a month writing a report), and things of that sort.

I think something like that, sort of mini-groups for mini-Manhattan projects. Of course, the whole thing would depend on the choice of people to run it. You have to have faith and you have to have quality in the individuals. They have to be absolutely honest; they have to be capable and sightful, and so on. Given that, to give a chunk of money to a group like that and say, "Go ahead and solve this problem if you think that you're committed to it and you can do it," is a different thing than we've got so far.

M. WEXLER: Are you referring, then, to some kind of research center?

ROBERTS: No, research groups. For example, three or four people in such a group might be operating out of different universities. As you remember, the Manhattan Project was delocalized, really. The production facilities and so on were [inaudible], but the University of Rochester, the University of Chicago, Columbia University and Los Alamos had branches. I don't mean anything that grandiose, of course, but as a pattern. When you're ready to make a push, damn it, make it and don't sit around writing contract proposals.

GUTHRIE: There's been much discussion at NIH about wishing to finance research per year with peer review. I'm sure you would like to make a recommendation about the length of time that you feel would be appropriate in giving funds to good research and researchers.

ROBERTS: If the judgment is sound at the beginning and adequately taken, I would like to see a review, certainly, at the end of three years. I think that the thing should go for a ten-year period if the problem is not solved. Of course, if it's solved earlier than that, that's the end of it. I think it should be a self-destruct situation. Nobody should be hanging on and saying, "I'm going to use this money to do something else." If it's directed for a purpose, it should self-destruct at the end, with the solution of the problem. That also is to be part of the package. I'd like to see a review at the end of three-year periods with a real commitment to a ten-year period if things are going well. I think that's what John Menkes was referring to, and not having these young people hanging on to a year-to-year thing with their

teeth and spending at least three-quarters of their time worrying instead of working.

M. WEXLER: You've had some rather extensive experience, I think, in connection with both Huntington's and schizophrenia and government-funded tissue banks. Would you say a word about that, Gene?

ROBERTS: I think that that's one of the most important things that can be done, to establish central sources of tissues and body fluids (like cerebrospinal fluid; blood; and in some cases, if people are interested, possibly even some urine samples, but that can get onerous and odorous). I think one of the severe limitations for a young investigator who suddenly gets interested in Huntington's disease (say some day at one of the meetings that Milt has organized) and who's really turned on is, "Where am I going to get the material? How am I going to ask those few questions that I want to ask to see if my idea is any good?" I think that the tissue banks could just serve a fantastic function, because then that person could write, "I would like to have three pieces of caudate nucleus from three controls, three Huntington's patients, three non-Huntington's, somebody with schizophrenia," or whatever, or "some cerebral spinal fluid, and I would like to do my thing on it and see if it's feasible." Then he would do it. But if he has to go scouting around for some neuropathologist to dig out a piece of tissue, forget it. When I've gone that route, I've gotten very close to zero material to work with. As far as Dr. Comings and I are concerned, we've gotten excellent cooperation and material from the one bank that we have here under Dr. Tourtellotte, and that's accelerated our work already. I think several such centers throughout the country would be extremely valuable, Milt. Really, a national resource of the first order.

M. WEXLER: Thank you very much, Dr. Roberts.

N. WEXLER: I just want to ask one question. I think that the problem you raised about funding is one of the problems we've been struggling with. As somebody said to our Commission, the government is notorious for being light on its feet, and it's very difficult to make a recommendation so that you have flexible funds.

One recommendation that was made was that there be a small grants program so that a young investigator could get approximately

\$5,000 just to try an idea to see whether or not it would be worth him going forward with, and the review process would be streamlined. One question is, what is your thought about that recommendation?

Another question is the problem of committing Institute funds for categorical programs. At the moment, the Neurology Institute is really suffering because it's extremely underfunded. It's committed to five current comprehensive epilepsy clinical research centers which cost \$1 million a piece. Every year there's around \$5 million immediately off the top for these centers, and those centers will probably increase. They have stroke centers and other kinds of categorical commitments. Somebody who wants to learn something fundamental about how the brain works, which could be applicable to Huntington's, Parkinson's, schizophrenia, across the board, is suffering. Some extremely well-known, well-established investigators, as I'm sure you both know, are having a terrible time getting funded through the Institute. Clearly, we need to have more funds for the Institute, but how do you juggle those two pressures?

ROBERTS: I think the first thing you mentioned is a sure tragedy. I was in Washington recently for something in the Neurological Institute, I think. Eight percent of the approved grants that are going...

...unconsciously is going to get preference over a young man. The person who's voting the priority just may be absolutely convinced that he's impartial, but he's going to be swayed. I just saw an instance of that last week at a meeting. It's just something you can't fight. I think that that's a complete and utter tragedy, because one-third of the funds of the Institute are now assigned to these categorical programs. My feeling is that there are only two kinds of science: good and bad. It's a shame when the really good things by young people, such as Nancy mentioned--undoubtedly, most of the breakthroughs are going to come from young people; the grayheads, they're the administrators--that they are being locked out. If only eight percent are being funded now and a third of that money is being diverted to these categorical programs, that compounds the tragedy.

The only way I can see out of it is to really lobby and push for enough money to fund all the approved grants, period. That's the way it was, you know, in those good old days. That's the way it should be because the reviews are fantastically rigid and the people are very critical. When something is approved by a

committee of 12 people, say, who are experts in the field, then it's likely to be worthy of being done. I think, the way it is now, you might just as well throw all the applications up that have been approved and pick up eight percent and just fund them. I think it's about that bad. There's no way around it, except to get enough funds to fund virtually everything that's approved. You have to fight for it, otherwise we're sunk.

With regard to the other question, I thought that at one time we did have this small grants program.

N. WEXLER: NIMH has it. I don't think NINCDS does.

ROBERTS: That's a great thing, to be able to give somebody \$5,000, or something like that, to buy the chemicals or the animals he needs, or to travel to a place where he needs to visit with somebody, whatever it is, so that he can try out the idea. If you don't have that, you can't move and you can't do it. I think that that program certainly should--everybody says, "Well, it's all more money, and everybody wants more money." How can I say that I didn't join that group just by talking the way I did. It's more money not just for the sake of more money, but it's more money facing the realities of the situation as they are today. It's such an utter tragedy.

I have a young man on my staff, a really fantastically bright young man, who has finished his postdoc and applied for his first grant. He came in in the third percentile over the cutoff for funding and got just a glowing report on his grant. He can hang that up on his wall, but he doesn't have the money with which to do his research. Several of us have to try to bolster his ego so that he gets in there and struggles for the next one. Completely unnecessary and so wasteful.

M. WEXLER: Sitting next to Dr. Roberts is Dr. Barklay, who is a prime example of what Dr. Roberts is talking about.

N. WEXLER: Even he should say that.

M. WEXLER: He has one of the most interesting, potentially creative research projects on the problem of Huntington's disease sitting at NIH, approved with a very high priority, and unfunded. I

don't know whether David wants to talk about that at all, but I wanted to mention it. Dr. Barklay?

BARKLAY: No, I don't.

M. WEXLER: Would you give your affiliation. Dr. David Barklay.

TESTIMONY OF
DAVID BARKLAY, M.D.
GRADUATE DIVISION
DEPARTMENT OF PATHOLOGY
UCLA MEDICAL CENTER

BARKLAY: I'm David Barklay. I'm a member of the faculty of the Graduate Division of the Department of Pathology at UCLA Medical Center. If you can't understand me, I've just come from the dentist. I'm never going to do that again before I have to testify before a Commission [laughter].

I'm going to try to be very brief, and I'm going to restrict my comments to two principal areas. First of all, I'm going to give you what might be regarded as a philosophical overview of where Huntington's disease stands in the world of science, and then I'm going to make simply one recommendation to the Commission as one area where its particular expertise might come to bear.

Dealing with any kind of a disease, and most particularly a disease like Huntington's disease, is extraordinarily difficult for a basic scientist such as myself. There was an HD Foundation workshop about a year and a half ago where a good friend of mine, Malcolm Steinberg, who's a professor of biology at Princeton, just threw up his hands in despair and said, "Look, there are times a real scientist studies a problem because he thinks the problem can be solved. Here we have a situation where we're dealing with a problem which may or may not be solvable, but the reason we're here is not because we know of any information which is going to lead to a solution, but because the problem itself is so important." The reason Malcolm would say anything like that is because of the way a scientist goes about doing his work.

We live in a world where we have a number of facts and they're related together in a logical order. We look at that, that body of knowledge and relationships, and we say, "Aha! Here, within this

body of relationships, is something that I don't know. But because of how I see the data and the logic fitting together, I think that I can use this information to find something new." He goes off, and he either succeeds or fails, having some idea as to where he's going to go before he even starts.

When you deal with something like Huntington's disease, you don't have that. Huntington's disease, at least as recently as two years ago, wasn't science. Huntington's disease was a human problem. You saw a person. You did not see a series of logical relationships which fit itself into the categories of test tubes and laboratory benches and white coats. If that's the case, why not give up? The reason you don't give up is because there are still tricks and there are still ways of doing it. One of the finest ways of doing it, and the way which I personally believe will succeed (if it hasn't succeeded already), is the kind of thing that the HD Foundation has been doing for the last five years. The point is this: A person who works on Huntington's disease for his entire life isn't the person you go to to ask how you cure Huntington's disease, because, clearly, that person hasn't succeeded. The people who have the information on Huntington's disease are the people who have never heard of it. The HD Foundation, by bringing in people who are interested in bacterial molecular genetics, or interested in biological clocks in the fruit fly, these are the people who might have the clues. Through workshops and through one device or another, what you do is you take Huntington's disease, or you take what your problem is and you try to lay planks between that human problem and the great body of logical relationships which already exists. You try to bring Huntington's disease into the relationships where science can do something about it.

Let me get just slightly technical for a moment. The easiest way to conceive of doing that, to most scientists, is to look for what's called a marker. When you look at a patient with Huntington's disease, you see a whole human being. In order to study Huntington's disease as a disease process (to find out what's causing it), what you do is, you try to take individual cells from that patient; you try to take just a portion, abstract away from the human being, the plumbing, if you will, and look at that and try to find the problem there. A marker is just a way of looking at the events in a test tube and seeing the disease process in those events in the test tube. Once you have that, the disease is part of science; and once you

have it as part of science, it's only a matter of time before you have a cure. It's possible, through the work of Gene Roberts, and I hope myself, and perhaps others, that such markers are either in existence or very, very close to being in existence.

I want to stop at that point and now perhaps shift just a little bit and mention something else which came out of the work of the HD Foundation, and that is something which initially was just part of a fundraising brochure, but it may be more than that: How do you justify studying a disease like Huntington's disease, which really affects a small number of people when you compare it with cancer, or heart disease, or something like this? The way you do that--and it's not just a justification, it's the truth--is that any disease is a window into all diseases. Huntington's disease is what's called a dominantly inherited disease. There are 400 other dominantly inherited diseases, and we don't know a damn thing about any of them. Some of those dominantly inherited diseases cause cancer, others cause heart disease. An understanding of Huntington's disease would open up vistas into an enormous number of other problems and would answer problems for which we haven't even had the wit to ask the question yet. Back to the marker.

I don't like to do this, but for just a moment let me refer to my own work. Huntington's disease, I started studying it to study Huntington's disease. Suddenly I find that quite by accident I'm also studying multiple sclerosis, and perhaps I'm studying a whole family of diseases known as the H-associated diseases. That's enough about the science.

All of this ultimately fits into what I regard as my recommendation to the Commission; that is, except for the people who are working in the institutes involved, there are clearly institutes at the National Institutes of Health that are overfunded. They're overfunded to the detriment of other institutes, such as the Neurological Institute. Superficially, the idea is a good one. You give more money to people studying cancer because more people are dying of cancer. But, in fact, what happens is that we don't know that we'll ever cure cancer by studying cancer. It may be that locked within the people who have Huntington's disease is the cure for cancer. You don't reserve all of your funds for a few restricted areas to the serious detriment of the other areas which have legitimate areas to pursue.

I guess my point to the Commission is this: What I've just described is not a new idea. It's one of the major political undercurrents which has been part of the National Institutes of Health long before Richard Nixon, but Nixon helped a great deal by re-establishing NCI. The point is, in a world of political reality we are not going to get that much more money into the NIH. The amount of money that is there is the amount of money that's going to be there in five or ten years. The problem is going to be, given that reality, how to redistribute the pie so that the most effective use can be made of that pie to solve human problems. I would recommend to the Commission that it join with the other voices which have been mentioning this problem now for several years; that it's not only a matter of the Neurological Institute feeling that it's been cheated because it isn't getting enough money, the Cancer Institute is being cheated as well because it is missing out on some vital information that some other institute could give it if it only had the money to pursue its own work.

Science is done largely by accident. You look to solve one problem and you solve another by mistake. The glory of science and the beauty of science lies in that. Gene Roberts said it very well: "Good science is done by good scientists. It is not done by administrators who decide X number of dollars here, X number of dollars there." Good science is done by opening the free market to good ideas and letting good ideas come to the fore and compete for the funds, as long as that's done fairly. That concludes my testimony.

GUTHRIE: Because I do agree with you, would you comment on the value of a large, broad constituency of health-related agencies all working together to pursue the opportunity as constituents of getting that money redistributed as it should be? That is really why Cancer got the money it got.

BARKLAY: I am a firm believer in the Milton Friedman view of scientific funding: You open the free market and you let good ideas and people with good administrative know-how come in and fight as hard as they possibly can for their piece of the pie. As long as there aren't rigid rules preventing fair fights in such a market, things will be distributed pretty well. So, yes. My answer to you is that you fight just as hard as you possibly can for as big a piece of that pie as you can, because others are going to be fighting just as hard in return.

N. WEXLER: I think Marjorie is asking it as a question. I'd rather make it as a statement. I'm not as pessimistic as you are about not having increases in the NIH. Mental Health got 50 million extra dollars this year, and none of the Institutes were cut, or not appreciably. It's going to be very hard to take money away from the Cancer Institute, or Heart and Lung. The people that direct these institutes are very well-placed and they're very close to the people in the White House who count.

I think, however--and I think Marjorie was implying this--that part of the reason that these institutes get so well funded is that they have one united constituency; that whether you have breast cancer, or brain cancer, or toe cancer, you're a cancer patient. Most people don't think of themselves as a brain patient or a gene patient; they have Huntington's, or they have multiple sclerosis, or they have muscular dystrophy, or whatever. The problem that we have is that there are an awfully lot of legislators in Washington who never hear from us. If all of these neurological diseases would get together and forget what our own special needs are for a moment, we are a tremendously powerful constituency. But we don't think of ourselves as having anything in common with that other person who has a neurological disease. I think that's one of the reasons why the Neurology Institute has been absolutely suffering abysmally, because they are either scientists or administrators and they can't get out and politick.

The Commission, in some ways, is a very political creature. The reason that we want to hear from you is that you really are the people who are going to keep on with the work of the Commission; that when the Commission is over in September, you're the people who are going to have to get on the phone and write letters to Congressmen to make sure that our recommendations for more money for this Institute to get people like Dr. Barklay, Dr. Roberts, Dr. Menkes, and Dr. Comings (from whom you'll be hearing) funded. I think that there can be more money, but it's not going to happen through a nine-member commission. It's only going to happen through people writing to the legislators, and they listen. One thing I've learned in Washington, which surprised me, is that they really do listen and they count how many people they hear from.

M. WEXLER: I have to begin to act like a chairman and be arbitrary and push a little bit for time. Thank you, Dr. Barklay.

I would like to call next Donald Ward, Dr. Phyllis Moeller, and Dr. Pieter Kark. If all three of you would come up, I know that all of you are under some pressure. We'd like to hear from you. I just want to introduce somebody who just came in to say hello. This is Bud Cort. Turn around, Bud, and say hello.

CORT: Can I say a word?

M. WEXLER: Fine.

TESTIMONY OF
BUD CORT
LOS ANGELES, CALIFORNIA

CORT: Hello. My name is Bud Cort. I don't know that much about Huntington's disease. My father had multiple sclerosis and died from it. I do know a little bit about schizophrenia. I am an actor [laughter].

I just want to say that Milton was my doctor and changed me from a frog into a prince, regardless of my appearance here today. Please support him, support his work. I do. I love him and love what he stands for. Thank you.

M. WEXLER: Thanks very much, Bud--prince. Thanks for coming down. Dr. Moeller, would you begin and give us your affiliation.

TESTIMONY OF
PHYLLIS MOELLER, M.D.
GENERAL PRACTITIONER
GLENDALE, CALIFORNIA

MOELLER: I'm Phyllis Moeller. I practice general practice in Glendale. I have no expertise, as Dr. Barklay does, in the Huntington's field. I got involved quite by accident through a friend (who is a social worker) who asked me--practically begged me--if I would take on a Huntington's patient. I have sort of an affinity for old people in convalescent homes for some reason I don't quite understand. I have dealt with people who have problems that a lot of people don't want to handle. On that basis I thought, "Well, might as well give it a try."

I've found the families most appreciative of any interest on this part. My plea here today is only for some funding for convalescent facilities for these patients. As was said by Dr. Barklay, we have human beings with problems more evident than the problem-solving at this point. I feel that the best way to handle the immediate problem is to have facilities that are particularly geared toward this treatment so that not only the personnel working in the facilities, but physicians who are interested, or who might be interested, can learn how to take care of these patients with their problems as they are.

I have had one rather difficult experience where the family, and the patient originally, had arranged to have brain tissue examined post mortem. I won't go into the details of the semi-legal arrangements that I made to make this possible when it happened unexpectedly, but I think it could have been facilitated. I think that more research material could be available in a facility where there is specific treatment for Huntington's patients.

GUTHRIE: Dr. Moeller, you're working with patients, and your recommendation is one that we're very concerned with. We've been discussing all kinds of care facilities. Could you perhaps describe what you think might be an appropriate care facility?

MOELLER: My only experience is with just generally oriented convalescent facilities. From that standpoint, I would think that relatively small facilities scattered throughout the community where the patient would be reasonably close to family for contact, where perhaps more physicians might be involved and become interested in Huntington's disease would be one recommendation. The other is that inservice training for personnel in these facilities is always required. In particular, Huntington's patients, as most of you know, can be very difficult. It's oftentimes frightening for personnel who are not familiar with Huntington's patients to deal with these people. If they are trained by a sympathetic, concerned physician in how to approach the patient--adequate medication is given to make it possible for the patient to be as cooperative as possible--these things can be worked out to the better care of the patient.

N. WEXLER: I was impressed by something you said in your response to the survey done by Dr. Barnard. You said that if there were adequate training in the care of patients and adequate

medication, HD patients would not need to be held in restraints. Would you just amplify that for the record?

MOELLER: Again, I fell into this quite by accident. I think I probably only used a medication called Haldol simply because it was relatively new and was recommended for helping patients with problems of a rather severe nature. So, I tried it. It seemed to work out very well with a patient that I had originally and with several patients after that. Subsequently, with one patient that was much more severe (was farther along), I found out that the management required medication early in the day, earlier than nurses are usually used to giving medication on a routine basis. So all these things had to be individualized to give large enough amounts in order to make the patient simply quiet enough musculo-skeletal-wise to allow them to eat breakfast, for instance, or to participate in bathing, or allow baths to be given to them. Does that answer your question?

N. WEXLER: Thank you.

MOELLER: I do think that sometimes physicians--and I don't know about any of my colleagues here--are reluctant these days to use any more than absolutely necessary: the minimum, minimum, minimum. We are under a constant constraint from the government not to oversedate patients in convalescent hospitals. In this case, I think one has to be a little willing to push it up in order to get the best effect and still not oversedate the patient.

M. WEXLER: Thank you, Dr. Moeller. Dr. Kark, would you give your name and affiliation.

TESTIMONY OF
PIETER KARK, M.D.
ASSISTANT PROFESSOR OF NEUROLOGY
REED NEUROLOGICAL RESEARCH CENTER, UCLA

KARK: Thank you. Pieter Kark, Reed Neurological Research Center, UCLA, which sums up what I do: research on neurological diseases, teaching about them, and a little patient care. I am not by any means an expert on Huntington's disease. My field of interest is closely related, though; and in view of the discussion that Mrs. Guthrie had with Dr. Barklay, it's with the inherited ataxias.

I gather that there is no blackboard, Mr. Chairman. I had a prepared statement, which you have. I wondered whether, instead of going through that, I might just read what I think are the four major points there and then expand on whichever or each of those points about which the Commission feels it perhaps does not already have sufficient testimony. I'm sure you've heard a lot of these covered by others.

M. WEXLER: Please.

KARK: First, my feeling is that in each patient with Huntington's disease the disease is very likely due to a single defective enzyme, or protein, whose activity is probably decreased by about 50 percent. A corollary to that is that there may be a small or large group of enzymes, each of which, if defective, could give rise to a disease that we would now classify in the broad spectrum of Huntington's disease.

The second major point is that I feel that there are firm possibilities of preventing Huntington's disease, if only we knew the affected enzyme or enzymes. As a corollary, I think it is also likely that we might be able to find some effective treatment if we knew this information.

The third point is perhaps a bit more controversial. My feeling is that to find the enzyme defect, there must be broadly based research on various aspects of basic neurochemistry, on various other inherited and toxic and neurological diseases, and on various basic neural mechanisms. The point could be made that just as Huntington's disease research could give rise to a better understanding of cancer, it's also likely that some other neurological disease might have to be understood before Huntington's disease itself could be solved.

Finally, my feeling is that this can only be done with broadly based, adequate funding of fundamental neurochemistry research, as well as fundamental research in the related neurosciences and of research at the interface between basic and clinical work. I have a little bit of information that I got from the National Institutes of Health in the last couple of days over the telephone. If it can be interpreted at face value, it would suggest that at the moment there is really quite a small proportion of funds going from NINCDS

into such basic research through the extramural programs as compared to money that is being spent on applied research and clinical work, and so on. I say that not to belittle the points that you made, because they're very important, but just to say that something else is also important and is being rather neglected at the moment.

Those are my four comments, and I'll be glad to go into whatever details the Commission feels would be appropriate.

GUTHRIE: Then I assume that you do agree that there is a great need for a lobby, if I may call it that, an educational program amongst families as to how we are related, how our diseases are related, so that we could get that broad constituency. I think we have to be very political, and that's why I'm saying it that way.

KARK: I believe I first heard you say that to me unofficially in Toronto about a year ago. My "yes" has not changed in that time.

GUTHRIE: Thank you.

M. WEXLER: I think your first two points are terribly encouraging. I'd like to focus on the last point, which deals with the problem of fundamental and basic research not necessarily directed at some particular categorical disease. I wonder if you could spell out (a) what kind of recruitment you think would be necessary to have people involved in that kind of fundamental research, or whether you think there are adequate numbers now; (b) what kind of funding program are you thinking of? Is it in terms of individual grant applications in this connection, or the establishment of research centers for this purpose, or something of this sort? Do you have a particular kind of program in mind?

KARK: Let me take the first question, if I may, first, Dr. Wexler; that is, how we're going to recruit people to do this. I don't have figures upon which to base what I'm going to say. But my impression in the 15 years since I entered and left medical school and been in academic medicine is that young physicians and young scientists coming out of graduate school (who are the future for all of the diseases we're worried about at the moment) tend to go, to some extent, where the money is. If they are interested in a field but there's just no way they can get a job in it, they're not going to do it.

About nine months ago, perhaps a little bit more, I went through an Affirmative Action Program in connection with hiring someone to work on diseases with me. I advertised for one month in Science, and I had something like 35 or 40 responses from extraordinarily well-qualified people from around the United States and even overseas, all of whom wanted this single job, and all of whom made it clear in their letters (directly or by implication) that they were not only competent, but that they were having a hell of a time trying to find a job where they could do research in basic neurochemistry or the borderline between basic and clinical research.

I don't think there's any lack of people. I think the problem is getting funds available so that people who are established can keep their labs going and can bring new people in to train them, and so that people who are young and bright and able to start on their own can start on their own.

Your second question was how should this be done? First, let me say that I think there are probably a number of ways that it could be done. Next, let me give you just a couple of figures and use those to illustrate what may be one of the problems. I understand that in 1975 approximately \$2.18 million were spent on fundamental neuroscience research at NIH (largely NINCDS, I would guess, though I'm not entirely sure of that point); and in 1976, \$2.63 million. In the same year the total extramural grants of NINCDS, based on a four-quarter year (as I understand it from a telephone conversation today), were \$87.4 million and \$87.2 million. I haven't done the calculation, but I would guess that it would be about three percent of the extramural funding that is going to basic research in the neurosciences, basic research which is absolutely critical for any neurological disease, communicable disease, stroke, whatever. We can't move forward based on what we know. In every instance that I know of where a disease has been tackled successfully in the last two decades, it has been tackled successfully only in connection with breaking some new barriers in basic understanding of biology, usually neurochemistry and the diseases that I work with.

I don't see anything wrong with research institutes in various parts of the country, provided they're not tied to one disease. It's a good thing to have people with common interests working together, even if it's an electroencephalographer with a basic geneticist with a basic neurochemist. They feed each other ideas, and they get good results that way. I think it would be very unwise to have a

single institute that dealt with a single disease the way muscular dystrophy had in New York a few years ago. That turned out to be really less than satisfactory. I know of no other instance where such an institute has been successful. The problem is that you have people scattered around the country with bright ideas, and you've got to get them working, wherever they are, try to get other people to go and work with them, or get them to work together at times. But you can't expect to pull them all together into one arbitrary city or one arbitrary center.

I think that one effective mechanism would be to shift the amount of money that is being spent on other things in the extramural programs of NINCDS into programs that are available for basic neurochemistry and neurochemistry clinical interface.

Another possibility--and I say this never having sat on a study section at NIH, so I may be wrong in my assumptions of what happens. I'm under the impression that in this time of very little funding there's a reluctance on the part of study sections and councils to approve and fund grants that are totally new and totally experimental in the sense of "Here's a new idea that needs to be tried, but as yet no basis for trying it." Yet there's very little money in the private sector to get people started to do their basic grants. I know because that's where I am. I've managed to squeeze some money out, but it really was squeezing. I would say I spent half my time trying to find the money to do the experiments that I would like to do instead of being able to get on with my job. A number of my colleagues are in the same position around the country.

It might be worth thinking about some kind of small-grant or partial-grant mechanism (such as I believe there is, or was, in NIMH) which might be a little bit more ready to try totally new things that sound good, that seem as though they have promise, with the idea of renewing them on an annual or two-year basis if they prove fruitful, increasing them if they prove fruitful, turn them into full-blown grants if they prove fruitful, but cutting them off if they don't. I think that's all I'll say on that point.

M. WEXLER: Thank you, Dr. Kark. The next witness is Mr. Donald Ward. Mr. Ward?

TESTIMONY OF
DONALD K. WARD, M.S.W., A.C.S.W.
BAY HARBOR REHABILITATION CENTER

WARD: I'm Don Ward. I'm a social worker at the Bay Harbor Rehabilitation Center, in Torrance. This is a new facility in the sense that we've just opened a large rehab unit. At the present time we are serving 180 patients. These are stroke and fracture patients, and we have one patient who has Huntington's disease. She is the reason I'm here. I wanted to talk about her progress. I would like to take this opportunity to introduce her, because she's in the audience. Betty, would you stand up, please, so we can meet you? Betty has consented to allow me to talk about the type of treatment and the work that we've done together.

It isn't just a one-person operation. I want to emphasize that it's a center. We have a nursing service, and we have occupational therapy, physical therapy, speech therapy, and social services, of which I am the only one.

However, Betty and I began working together about a year ago. We work as a team at the Center. We operate under a system called PPG, which is not the Pittsburgh Plate Glass; it's Problems, Plans, and Goals. This is the team approach that we've used, everybody, including our volunteer services, and it has made a big impact on whatever progress we've made. To illustrate how our volunteers have contributed, one of the volunteers (and one of Betty's good friends) brought her up here today so that she could be with us and be a part of the program.

When Betty first came to the center a year ago, it was because she'd had some unhappy experiences at another facility, and she had decided she needed a change. She came to our facility, and we were a little bewildered to know just how to approach this because she was the first and only Huntington's disease patient that any of us had met. At that time, Betty was not talking, and she was not walking. She was having troubles of isolation from her family, and many of her rights had been taken from her. These were the problems. These constitute the problems that we saw as a team when Betty came to the Center.

What we did, in terms of our treatment, we began to work on the Plan stage. We had isolated and identified the problems. What we

had to do as a team was to come up with some plans and something that would help Betty. She was highly motivated at that point to think that there were some opportunities for her, so she cooperated in everything that we did. We started first with her speech problem, and we had a speech therapist work with her. It turned out that the only problem she really had was that her speech was too rapid. She spoke so fast and so rapidly that the words ran together and she couldn't be understood. Since the speech therapist had given us this insight, Betty practiced and worked with a speech therapist for several weeks. At that point, she was beginning to speak very clearly. She could say what bothered her. She could talk to her doctor. She could do all of the things that she hadn't been able to do.

She couldn't walk when she came. Through our activities program (our restorative, occupational, and physical therapy) Betty began walking. At this point, she's doing an excellent job. She doesn't need any assistance. She walks by herself. She takes care of all of her personal needs, and she's handling many of her own financial affairs. As the program developed, and as she began to speak and to walk, then she was able to develop contacts with her family which had been completely isolated from her. The family and many of her friends come to visit her, take her out of the facility to activities off the grounds and into the community. Betty has now developed the contacts with church, with the friends, and with the family that she'd been missing for the past year.

The last thing, I think (and most important, maybe, in some respects), is that the patient's rights had really been violated. At this point, Betty's in the process of hiring an attorney who is going back to court with her to establish the rights that have been taken from her and to get her back to an independent state, as near as we can, in a board-and-care facility, hopefully, or perhaps in her own apartment in the community.

This is really our system of the Problems, the Plans, and the Goals. Betty, thank you for allowing me to use this illustration.

HILL: Thank you for doing it.

WARD: I think it has demonstrated that there are many things that can be done right now in the community without really any additional expense. Much of our program is already federally

financed through Medicare and MediCal and private patients. I think we're having an ongoing program in one of our local communities that could serve as maybe a goal or an example of some of the things that could be done in the community.

We'd like to issue an invitation to the Commission, if you'd like, to come and visit the Bay Harbor Rehabilitation Center (at Torrance) and see if this might be something that could be pursued in other communities. Thank you.

M. WEXLER: We certainly appreciate that, and it's a remarkable illustration of what can be done. I think individual Commissioners would certainly like to visit. I wonder if I could ask if you have any kind of literature at all that would explain your program and explain the facility. We'd love to have that.

WARD: It's just being developed. We don't have it finished. One of the things we're waiting for is to complete the outpatient program, which hasn't been certified. Once the outpatient program is certified, we will be getting out more literature. There is some, however, and I'll be glad to send that to you.

M. WEXLER: Thank you very much. Thank you, Dr. Moeller.

MOELLER: Could I just ask a question? Would you have, or did you have any problem in getting MediCal certification for the rehabilitation services which are usually more expensive, obviously, than an ordinary convalescent facility?

WARD: We've just finished the program of supervision. The Department of Health has just certified the program as one of two rehab centers in Los Angeles County. When we complete the outpatient physical plant, the next step will be to get certification for the outpatient program. So far, it is certified as a rehab center and a skilled nursing facility by the State Department of Health.

GUTHRIE: One question. First, let me commend you. I'm just thrilled to hear what you're saying because I've been doing a lecture called "The Team Approach to Huntington's," and it goes along very much with what you're saying. I'm interested in asking, how does Betty feel, would you know, about her relationship to (being the patient with Huntington's disease) and living with, I

assume, other people who have other problems? I'm interested in that feeling that she may have.

WARD: Let me tell you a little bit about the approach that we used with Betty, with her consent. In order to come out of this stage of depression and to get involved, we had her come into many of our activities programs. At first, of course, she didn't participate as much, but lately she is very active. She has been responsible for working with other patients who have been isolating themselves to get them out of their rooms and into programs. She's become a leader, in a sense, with the other patients by example, I guess, as much as anything. The fact that she could overcome and do as well as she did with so many handicaps has been an inspiration for a number of other patients.

If we could, and we had enough Huntington's disease patients at our facility, what we'd like would be to have Betty as a group leader--she has expressed an interest in this as a further part of her treatment--have group sessions and have Betty as one of the leaders. She would enjoy this and I think would be the kind of a leader who would inspire other Huntington's patients to do more for themselves.

M. WEXLER: I would just like to get a brief impression. In your estimation, would it be advantageous in this kind of program to have groups of patients with mixed diagnoses, or would you think it more advantageous to have a collection of HD patients?

WARD: We do have some other patients with other diagnoses, multiple sclerosis. I think all of these related-type illnesses could be included in groups; they're similar. I think the depressed state, the feeling of worthlessness, and the feeling that they can't do anything can be overcome by the group approach or the group leader who has already overcome the problem.

M. WEXLER: Thank you very much. Mr. Fritts, Mr. Commerford, and Dr. Cohenour are up. I'll be right with you, but we have a very brief presentation. I want all of you to meet Judith Bradford Rini, Dulcy Bradford Rimac, Mary Ellen Bradford, and Ethel Bradford. Would all of you come up, please? I'll let you choose your own spokesperson or the order in which you're going to speak. Please introduce yourselves.

TESTIMONY OF
JUDITH BRADFORD RINI
HUNTINGTON BEACH, CALIFORNIA

RINI: My name is Judith Rini. This is my sister Dulcy, my grandmother Mrs. Bradford, and my mother Mrs. Bradford. We're here to talk about the fact that my father died of Huntington's disease a year ago in September. We're all very nervous. I know there would be a lot more HD families and patients here if it weren't for the fact that you feel a little uncomfortable sharing sorrows and feelings and traumas. We want to share our feelings.

I'm an at-risk person, as are my sister and my younger brother. When I talk about it a lot of people react to this as, "This must be very traumatic," but our family has a very positive attitude towards HD. In fact, it's made us appreciate life and do all the things that we might have put off, so it has had a very positive effect. Really, the tragedy to me was to see my father, who was a very proud, brilliant history professor, suffer for many, many years and be ridiculed because no one knew what he had, what was wrong with him. We thought he was having a nervous breakdown, et cetera. He went to many doctors (neurologists, psychologists). He went to every type of doctor, with no diagnosis. Ultimately, he was finally properly diagnosed after many years. This doctor (neurologist) who diagnosed him prescribed heavy sedatives, so for at least two years--it may have been three years--he was drooling and nodding and couldn't participate in life. He was terribly depressed and ultimately attempted suicide. He did not succeed.

Soon thereafter we found out about CCHD. Through them, we found Dr. [Selleck], who did a miraculous thing: he treated my father as a human being rather than as a guinea pig or an HD patient. He gave my father hope and the proper medication, and my father was, in essence, reborn and lived on several years thereafter with a very positive attitude. That's what I want to say. Being at risk isn't nearly as bad as seeing what happened to my father.

I think that doctors need to be educated, society needs to be educated. When we said, "HD" (because I saw that my aunt had it), our M.D. said, "No, can't be that. Only men get HD. Women never get it. It's only in one family back East," and so on and so forth.

TESTIMONY OF
DULCY BRADFORD RIMAC
VALENCIA, CALIFORNIA

RIMAC: I'd just reaffirm a couple of things she said. For me, I've made very careful plans for my life. I realize that plans need to be flexible, but it's put a very clear time line in my mind. I'm going as quickly as I can to make the most of the years I have. In that essence, it certainly is depressing and there are problems associated with it, but it's not debilitating for me. My father's life was a short one but a very rich one, and that is the example that we follow. For me, it will not be so hard because I will know what's coming. That was the biggest problem. The biggest obstacle is the unknown, always. For us, things are better.

I have two children and hope for them that things will be better yet. That was my assumption when I had them. One of the recommendations our fine neurologist made was that we all ought to be sterilized. That was the first thing he said after he announced the diagnosis. That, I think, was not his place. That was something that we all had to come around to. It was like a slap in the face. I think he could have referred us to someone else. It would have been more appropriate than attempting it himself, because he was just not a compassionate person and was not interested in us as human beings; rather, statistics and examples. That's the way he dealt with us, and I think that was wrong. That was the really difficult thing we had to face in this.

TESTIMONY OF
MARY ELLEN BRADFORD
GARDENA, CALIFORNIA

M. BRADFORD: As you can imagine, I'm very proud of my children. The surprise and the shock of my husband's illness was quite amazingly difficult. He was such a brilliant man that it masked the illness, I think, for quite a long time. Because we didn't have a known family history of it, we weren't expecting it at all. He went to doctors often, as Judy said, and the family history is what threw them off all the time. Don's mind was so excellent that it just wasn't in the list of things to be worried about.

When it came, it came very coldly, a tremendous shock. A doctor told me in a hallway, "Well, your husband has Huntington's

disease," and I almost fainted. I knew enough. I had done enough reading to know that that might be. When it came, it was so cold. This first doctor we were sent to was almost cruel in his attitude, his behavior. We went from kindness, where people were trying their best, to this terrible brick wall. My children have been magnificent. They say, "Quality, not quantity"; in other words, their lives are going to be the best. We've been able to handle it that way.

I am so thrilled to meet you people on the Commission and to meet these doctors that I'd heard about and that are in my prayers every night, names that I've grown to think the world of. Your work and your optimism in those early meetings when we first met have kept us all going. We're proud to be a part of a very exciting brink in medical history.

M. WEXLER: Just one simple question. You had your children before you knew anything about this kind of threat in your family?

M. BRADFORD: Yes. We only found out in 1969.

RINI: I'd already had my first child by then.

M. WEXLER: There was already a grandchild?

M. BRADFORD: There was already one.

RINI: Now there are four.

TESTIMONY OF
ETHEL BRADFORD
HERMOSA BEACH, CALIFORNIA

E. BRADFORD: I'm the mother of Professor Bradford. I lived with them in his last years because she was working, so I had personal contact with him daily and almost hourly. We would take walks every day. I thought that kept him going a lot. He loved it. What bothered me (and now I know that that is not true) was that I felt that people didn't care about this man; the public and, above all, the doctors didn't care. They just acted as though it was something that either didn't exist or it would go away. That kept coming to me.

To me, the saddest thing has been to deprive the HD patient of his dignity and their sense of well-being. I have a daughter with the same affliction who lives in my home with me. She has two teenage children. Their father is dead. The children would not come. They are 17 and 19 years old. They are trying to act like it doesn't exist, because they know it's hanging over their heads. All they've ever been told is not to plan on getting married, or having a family, or leading a normal life.

When Mrs. Guthrie got this going, I found out that people care. They didn't know before. To me, that is the most moving thing, that there is hope. What this man said, who has this place in Torrance, I think that that is wonderful, that they can have these people and give them hope and find out that all is not lost. That's all I have to say, that I have found that people do care. Thank you.

M. WEXLER: Thank you. I think the existence of this room and these people indicates how deeply and how profoundly they do care. Thank you for coming.

E. BRADFORD: Thank you.

M. WEXLER: I would now like to call on Dr. Markham, Dr. Cohenour, Mr. Fritts, and Mr. Commerford. Could we hear first from Dr. Markham. Would you give us your affiliation and title, Dr. Markham.

TESTIMONY OF
CHARLES MARKHAM, M.D.
PROFESSOR OF NEUROLOGY
UNIVERSITY OF CALIFORNIA, LOS ANGELES

MARKHAM: I'm Charles Markham. I'm a professor of neurology at UCLA. I'm a combination physician and scientist, I'd guess you'd say. I've been working with the problem of Huntington's chorea as with other--what we call basal ganglia diseases for some years' time as a physician and have also participated in a variety of research endeavors to try to find out something about the diseases in an effort to hopefully cure it one day.

I speak largely as a scientist, and I'm fully appreciative of the problems of the patients and their families and their many friends. But because of my job and my interests, I'm looking more to the cause of the disease and what we can do about it. In other

words, the treatment that we can present; we can deal with it on today's level is essential for today's people. But I would say we must look 10 years, 20 years, and a further number of years into the future to really get to the bottom of this problem in a curative sense.

How do we arrive at such a cure, at such a point of first-rate treatment? As I see it, it has to be a combination of research on the one hand, and education and treatment on the other. These things sound sort of separate, but really, they're not. They are virtually inseparable. Again, I would say the first of these is research to try to find out what is going on in this disease. I've tried to work out in my own mind (and also have given you a summary of my thoughts) what might be called kind of a flow diagram that would allow us to tackle this particular disease and to get to some kind of a solution for it.

First of all, there are two main research efforts that have to be taken, each one will have a kind of a stepwise, progressive followup. Perhaps the most important aspect of research is what is causing the disease at a cellular level? There is a deficit of something, an enzyme, a particular biochemical structure. We don't know what that is yet. It's crucial for us to identify this particular substance, due to the lack of which people in their typical middle years will have a failure of many cells in their bodies. The trouble is that the people who are the most likely to find this basic defect are also scientists. I'm talking about the scientists who don't know very much about Huntington's chorea. They're scientists who deal with cell biology, membranes, proteins and other structures inside cells. They are often as ignorant as the average layman as to what Huntington's chorea is. One of the jobs is to introduce the problem of Huntington's chorea to these scientists so that they, in turn, can have their thoughts and their work stimulated.

Once we find the particular chemical deficit, there are a series of steps that can be taken to treat the disease. In other words, after we find out certain aspects, then the treatment comes next. It may turn out that the substance that is lacking can simply be given as a therapeutic substance, maybe something like giving insulin to diabetics, maybe something like giving dopa to people with Parkinson's disease. It may be that the step will come in a different way. Maybe this substance, whatever it is, will be able

to be detected in utero in women who are pregnant. If this were the case, this would lead to a possible prevention of a newborn who has the disease. Supposing it could be identified by using a slender needle placed in the woman's abdomen and a little bit of the amniotic fluid taken out and having those cells in that fluid analyzed? Supposing she could be told, "Your child doesn't have Huntington's chorea," or, on the other hand, she was told, "Your child is going to get Huntington's chorea one day." She may well choose to have an abortion at that stage. This problem has been faced in an illness called Tay-Sachs disease. This can now be identified by these amniotic fluid samples. There's a good chance, in a few generations' time, that this illness may be wiped out completely. If this kind of substance can be detected in Huntington's chorea, this would be a major step ahead.

The other major push--I'm talking now about what is another major research push--is to look at those areas of the body that are hardest hit by the disease and learn more about them, so that we can treat or prevent the symptoms from developing. This won't prevent the disease, but it will allow us to get several steps ahead of the disease, if I can put it that way. If we look at a part of the brain called the neostriatum, the caudate nucleus, and putamen (which is a factor in many of the movements that one sees in Huntington's chorea) it's quite possible that if we know more about this area, we'll again be able to give a substance that is in this area. There may be too much of the substance, in which case we can reduce it; or of there is not enough, we can give it artificially. This may sound a little like what I was talking about in the beginning, but it does represent a fundamentally different approach.

Thirdly, is the matter of education. There are two main groups to be educated: The patients and their families are one group, of course. It's surprising how many people with the disease are just finding out about it today. I look in this audience and I'd say these are the knowledgeable people, but we have to get to the people who are not knowledgeable. This is going to take a lot in the way of publicity. Also, it has to be made very clear what the genetic implications of this disease are. Today, the only thing a person who is at risk can do is to not have children. As hard as this fact is, they have to be given the opportunity to weigh this choice themselves: "Shall I or shall I not have children?" If they don't know, they don't have a choice, so they have to be given a choice.

The other group that has to be educated are the scientists who can help us find out something about this disease, but who don't know anything about Huntington's chorea. As I mentioned in the beginning, the scientists that are probably liable to help most in this disease are the ones who don't know much about it right now. I would say that through the efforts of the HD Foundation in having small workshops of scientists, in introducing those who have some clinical experience (such as myself) to those who have some basic knowledge that might be the real tool to solve this disease, to bring us together repeatedly, there may be some hope. As I see it, it's only by this route of finding the right scientists with the right ideas at the right time that we're going to see a real solution to this disease. Thank you.

M. WEXLER: Thank you very much.

GUTHRIE: I just want to comment that you left out somebody else that has to be educated, the family physicians. We feel very strongly about this.

MARKHAM: Yes, I know that.

GUTHRIE: I know that you know it, and I know that you just left it out. Another thing I would like to comment on, which you might like to know, is that with all the recent publicity, last month the Committee to Combat Huntington's Disease found 179 new HD families. Our average has been 33 a month. You are right. If we could get the word out, maybe we can find more of the people who are still not knowledgeable.

MARKHAM: Yes, you're certainly right. Many physicians don't know, and I think this is something where we physicians who do know can do a little better education job.

M. WEXLER: I want to say just for the audience that Dr. Markham is one of the longest time, earliest researchers in HD anywhere. He's one of the very early members of the HD Research Group connected with the World Neurological Organization, whose membership includes researchers from around the world. They come from Germany, Australia, Japan, and everywhere. In Dr. Markham's testimony we have one of the most knowledgeable of all physicians and scientists.

I wanted to ask you, Dr. Markham, aside from the workshop program, isn't there a need for some kind of scientific information retrieval and exchange that would keep people up with the latest developments in this kind of research? Do you have any suggestions as to how that should take place?

MARKHAM: I think you're right. There is a need in this regard. Again, I think we're dealing with two groups of people: one is the people who have the problem, or think they might, or know somebody who does. Disseminating information from them can take place, I think, through such fine organizations as yours and Mrs. Guthrie's. In terms of reaching the scientists who might do something for the disease, there are a number of possible ways that information can be spread. For example, there's a group located at UCLA called the Brain Information Services. It has a mailing list of between 15,000 and 20,000. These are scientists in the basic areas I'm speaking of and in related areas. There are many bulletins that go out (some go out every two weeks) covering a variety of subjects. This is a great source. There are other good information services, too, but this one happens to come to mind.

M. WEXLER: I'm going to ask a far-out question. Do you think there's any point in having a journal that deals basically with basal ganglia disorders, not necessarily an HD journal?

MARKHAM: I don't think so. I think it would reach too limited an audience. I would be trying to attract people to become aware of this disease who don't know much about it and who wouldn't automatically turn to that journal. I would like to see word of Huntington's chorea get in a journal such as Cell Biology, or Journal of Clinical Investigation, or Neurology, or Brain Research, or Experimental Brain Research. These are the kinds of journals that maybe somebody might read and say, "Oh, that's kind of interesting. That's in my area." One has to kind of bootleg articles into those journals, but that's what I would want to do.

May I say one more thing? I forgot to mention funding. Of course, funding in these problems is always an enormous challenge. However, it seems to me that within NIH there is a considerable amount of money for fixed expenditures that are very difficult to get around. Specifically, there are very large clinical units at Bethesda, Maryland, whose expenses go on year in and year out.

Sometimes they do superb investigations, but sometimes they're treading water. There are even moves afoot to build another institute there. Two years ago I heard from one of my esteemed colleagues at NIH that there was a plan to build a \$100 million hospital. I asked, "Why in the world should this be done?" It was said, "Well, we have to have an image. We have to alter our image." A \$100 million image is a pretty high price to me. I would divert money from certain fixed expenses within NIH and put it more into the grants and contracts end.

N. WEXLER: Would you make any specific recommendations for alterations in medical school curricula or in continuing medical education to cover HD more adequately?

MARKHAM: This is a spotty field, varying from medical school to medical school and class to class. It so happens that often medical students in their first and second year (when they're learning anatomy and some of the other basic subjects) will, in fact, see a patient with Huntington's chorea. The patient is brought in to illustrate to them a problem in a particular part of the brain that they are now learning about when the disease is related to that particular part of the brain. But that medical student could just as well go all through medical school without even hearing about it; so, it's spotty. In a word, if that could be corrected, it would be highly desirable.

M. WEXLER: Thank you, Dr. Markham. Dr. Cohenour?

TESTIMONY OF
ROBERT COHENOUR, M.D.
NEUROLOGY STAFF PHYSICIAN/PHYSICIAN COORDINATOR
NEUROSPECIMEN BANK OF AMERICA
WADSWORTH VA HOSPITAL
ASSISTANT PROFESSOR OF NEUROLOGY, UCLA

COHENOUR: My name is Robert Cohenour. I'm a neurology staff physician at Wadsworth VA, and have an appointment as an assistant professor of neurology at UCLA. My role vis-a-vis Huntington's disease is as Physician Coordinator of the Neurospecimen Bank of America, which is located at Wadsworth. At this point, we are the only full-service banking facility, you might say, for neurologic and psychiatric tissue in the United States. Within the last year

or so we have been given authority and funding to solicit tissue for the bank from Huntington's patients who have either died with the disease or were willing to provide blood and spinal fluid specimens, and also from their families. We're actively engaged in this project, and at present we have a large number of Huntington's brains in the bank, as well as spinal fluid and serum, which are to be made available to appropriate investigators in the field.

The reason I'm here is just as a general informational point and also to emphasize the need for a bank, at least one. My hope would be that eventually there could be regional banks serving numerous projects of this sort. The HD Foundation and CCHD have been preeminent in encouraging this sort of facility. I think that we're really on the threshold of a newly developing area. Today it's very difficult to obtain any type of tissue for research from human subjects, and to obtain tissue from a group of such subjects with a single disease or risk of a certain disease is even more difficult. Preparing this tissue under controlled circumstances and having it available to investigators at a reasonably short notice is another major reason for a bank such as ours. I think it's going to be absolutely necessary to have this type of facility available in the future so that reasonable research can go on with control and disease tissue, whether it be from living patients who have been willing to donate spinal fluid or blood, or whether this comes from post mortem tissue at autopsy.

I think eventually, if we could decentralize a little bit and have regional organizations, this certainly would facilitate research in various areas of the country. Right now we provide tissue from certain areas of the brain (and also the spinal fluid and spinal cord) that's prepared under specified circumstances to investigators who've been directed to us through the HD Foundation and CCHD. This is a major task in this day and age, with legal problems and the difficulties in obtaining autopsies, the general waning of interest in autopsies in teaching centers. One of my reasons for coming was to let all of you know about us and spread the word around a little bit to your contacts regarding the need for continued support of a banking facility. The Drs. Wexler and others who I have had contact with have been supporting this from the start, and I think we can continue to provide an improved service in the future.

We're always in need of additional tissue, primarily because there are certain areas of the brains from affected patients which

are of the greatest interest to investigators; and these, of course, become depleted most rapidly. We also maintain a facility for control tissue of all sorts and a coding mechanism so that investigations can be done at the highest level. I would like to emphasize that you can contact me or the Huntington's disease organizations (the HD Foundation and CCHD) if you know of anyone who is willing to meet with us to donate blood and spinal fluid, or who may be willing to donate post mortem tissue (some family member who may be afflicted and near death, and so forth). The need for this is overwhelming, I think. We will have to continue to be very solicitous, but also very straightforward about the necessity for having this sort of service available so that investigations such as those that Dr. Markham has been involved in for many years can go forward.

M. WEXLER: Thank you. I'm not really asking this question, I'm making a statement. Obviously, we're not going to get anywhere with a solution to the problem of Huntington's unless we have tissue to examine. Research is impossible without some kind of a tissue bank.

I want to ask you something more specifically and more particularly in connection with your suggestion about regional banks. I wonder if you could tell us some of the difficulties that exist when there is a bank only in one place, like California? For example, what about the problems of shipment, collection, et cetera?

COHENOUR: I think it's like having any other type of banking facility. The neighborhood bank serves a community and has a ripple effect in the areas around it; but, naturally, its greatest impact is locally, I think. It takes a great deal of communications efforts to spread the word to more distant locales. Of course, with the HD Foundation, this is not quite as big a problem as if we were trying to advertise ourselves, for instance. The banking facility acts as a catalyst from several points of view. If a person has a good idea as far as a research project, his biggest chore is to try to obtain appropriate tissue, and particularly enough tissue from enough individuals. With a bank there, this initial hump of obtaining the tissue can be overcome relatively easily. Also, there is feedback between the bank and the investigator. Again, proximity is influential, just knowing the people and being able to talk with them and not having a three-hour time gap between when they go to work and when the bank may go to work, for instance. These are day-to-day problems, but I think they do get into the picture.

Any one bank, naturally, is going to have a certain amount of pressure on it at any given time for orders of various sorts. Inevitably, I think to provide the best service there needs to be several similar organizations which can then exchange tissue. For instance, we deal with the coroner's office in this city. We have an unusual liaison with them in that California is the only state in the Union which has a law stipulating that tissue obtained for legal coroners' cases can be dispensed for research purposes without additional consent of the family. This sort of thing is really very important because most of this tissue is going into bottles and never will be used for anything. This kind of liaison should be established with coroners' offices in all the major cities.

A few weeks ago we had a letter from a prominent investigator (from Chicago) in another area who had been trying for something like six months to obtain one brain from the coroner's office or Cook County Hospital. The coroner was just adamantly against this type of use for the tissue. These are problems which we can help people with because we're familiar with what has gone on in order to get this changed in California. Our coroner is much more progressive, I'm afraid, in this situation. A liaison there, I think, could help establish a regionalized service.

M. WEXLER: Thank you, Dr. Cohenour. Again, I want to exercise a prerogative as chairman and say that we're going to be compelled to transfer our dinner hour from 5:30 to 6:30, unless somebody has some monumental problem about that. Let me know, or let this gentleman back here know about it, and we'll call on you earlier. I'm doing it in order to be able to accommodate several people who need to testify very shortly and be able to leave. If you have a real problem and have to leave, and this shift of dinner hour makes trouble for you, please advise this gentleman. I will then be able to call on Dr. Giordano, Gayle Soergel, Gene Bartolotti, Ruth Krueger, Dr. David Comings, and Lyndon Jerry in the period between now and 6:30. I hope to get them all in. We're a little behind time, and I want to catch up a little bit. Hopefully, this will not discommode anybody. We'll get the people I mentioned in to testify, and then we'll have dinner at 6:30. Ted Fritts, please. I'm going to ask you to give your full name and background, please.

TESTIMONY OF
ALFRED T. FRITTS
EXECUTIVE EDITOR
THE BAKERSFIELD CALIFORNIAN

FRITTS: I have little scientific contribution to make. I'm Ted Fritts. I reside in Bakersfield, California. By profession, I am a journalist, and I'm Executive Editor of The Bakersfield Californian. My reason for being here, although I am a journalist and recognize the need for the dissemination of material and educational material regarding Huntington's and other related diseases as part of my daily responsibility, I'm also an at-risk patient. My father died in 1969, not with Huntington's, but had been diagnosed some 15 years earlier (when diagnosis was very slim and it was given another name at the onset of his illness). At an appropriate time, when enough material and information was known about the disease, it was given the name of Huntington's. I have an aunt who has also been diagnosed and is currently residing in a convalescent home in Colorado.

I am 30 years old. I can certainly empathize with the two ladies who testified earlier as to some of the difficulties that they have encountered. Unlike them, I knew what my father had at the young age of about ten years old. I at least was knowledgeable that he had a disorder that had been recognized as a neurological defect.

Where it is difficult right now in my family environment is that as I come to the age where onset may be inevitable, I also recognize the problems of coping with daily life and have an additional responsibility to my brother who is 40 years old and who feels the pressure daily. Let us deal with some intangible problems with the mere knowledge of the disease. Mere knowledge of a disease, whether it be diabetes, or whatever, can afford some particularly difficult complexities in dealing with daily problems in that you may manifest in your mind particular symptoms of the disorder which have not been professionally diagnosed. Anxieties can compound, and almost everyone hates the uncomfortable feeling of undefined anxiety. If you can give it an answer or a reason for being, it becomes a convenient crutch.

If you have Huntington's in your family (and you're knowledgeable of it) and you should awake one night, jittery from sleep,

"My God," you say to yourself, "that is a symptom." How many of us have fallen down stairs and thought that we were blind? It's an incredible psychosomatic difficulty and should indicate to all of us the incredible amount of importance of prediagnostic treatment, of professional psychiatric or psychological counseling that can allay some of these fears, where we automatically try to assess our problem given some absurd symptoms that we automatically attach to a non-existent problem. I used that jerking in sleep because we all have had that sensation of being abruptly awakened and not really in a sense of consciousness, so we assume we have some problem. The awareness of the problem in itself provides a certain amount of irony to some of the problems. It can compound the problem.

As in the case of Betty, if I can refer to her, these anxieties can also compound to a point where you can indeed have a problem that is not there. As mentioned by the two ladies who are also at risk, there is also a positive feature to the illness. By knowing that I have a 50-50 chance of coming down with this disorder, my life is still disordered, but I am probably enjoying it just a little bit more in that I have some goal to look forward to. I use those terms intentionally. I look forward to working toward something whether I get it or not. What might foil it for me is that I could be hit by a taxicab upon leaving the building this evening.

Dr. Wexler explained in a letter to me the "cloud of anxiety." This is the toughest thing with this type of disorder. If you are dealing with another genetic disorder which may be in your family (hemophilia, diabetes, or other disorders), at least you are physically and mentally attuned to the possibility. This is not necessarily a physical disorder--and I speak out of school here--but it's compounded by the fact that it is a mental inclination as well. How do you deal with the problem in a daily life when you know the mental intrusion can impede your thinking? This is a cloud, the most difficult aspects. Activity seems to be the best remedy. As friends remark to me that I exhaust myself by spreading myself thin, the greatest virtue of my daily existence is the fact that I keep busy, that I have those who love and care for me surround me. This is the best type of facility provided, outside of a formal facility for rehabilitation such as being developed in Torrance.

There is a great hope that exists, evidenced here by the professionals who have testified and who are doing research in

the area. Those that are personally and professionally involved with the disorder are lending their time, effort, and money in making the general public aware, as well as the Congress and the general body of lawmakers. The breakthroughs have been phenomenal, not necessarily just for the layman, but the professional, who can see just a little spark in such a vague area. It ignites further curiosity, further excitement and enthusiasm. This is what keeps us all going. Whether or not it means that it can cure us who are of substantial age, it means hope for our children. It means hope for arresting certain traumatic elements of the disease and making our lives more comfortable, which our fathers and other relatives were unable to experience.

From a political argument, Congress must be made aware of the tremendous amount of cost of treating this illness. A history professor has been taken away from his function. He has provided instruction and education for hundreds and thousands of students. To be chopped down early in his life without the exercise to fulfill his obligation to render to thousands of students education has caused you and I and society a great loss in dollars and in talent. This can be compounded by the hundreds and thousands of individuals who suffer from this disorder. In real dollars to treat this illness it is difficult to fathom. My father was isolated within a very expensive facility for over ten years of his life. He also did contribute to the economics, to society and to all of us. It is a shame that we have to...

TESTIMONY OF
WILLIAM J. COMMERFORD
INSURANCE BROKER
REGIONAL TRUSTEE, NORTHERN CALIFORNIA CHAPTER
COMMITTEE TO COMBAT HUNTINGTON'S DISEASE

COMMERFORD: ...I originally met her and her husband in a social dancing group and enjoyed dancing with her very much. I knew her as a very warm, wonderful, loving and beautiful human being. Although her husband is a medical doctor and a psychiatrist, when she first began manifesting the early symptoms he did not recognize them. They went from neurologist to neurologist, and none of them could put a name on it. Eventually, a neurologist in Oakland, California, did. Her case, these last years and months in a hospital bed and in her own living room, illustrates, in my

mind, the very critical problem that Huntington's disease patients and their families face; that is, the problem of care.

My recommendations to the Commission are really in three areas: one is in the area of insurance (which happens to be my own field of expertise), and the other two are in the field of patient care. The problem with insurance is that commercial insurance policies (Blue Cross) and group insurance policies do not provide extended care, real extended care. They provide some basic coverage, perhaps 60 days of hospitalization, followed by 30, 40, or 50 days of what's called extended care. That's not very extended, certainly not for an HD patient who is going to need months or years of extended care.

Medicare is available to persons over 65, but it is also available to another class of people. Persons covered by Social Security who suffer from a permanent disability are eligible for Social Security disability retirement after five months, I believe, of disability. These people, then, though they may not be 65, are eligible for Medicare. HD patients would be eligible for Medicare if they have been covered by Social Security. The problem with Medicare is that there's something like 50 or 60 days of basic hospitalization and another 50 days of so-called extended care, and that's totally inadequate.

My recommendation is that the Commission urge Congress to enact legislation that provides true extended care through Medicare. Secondly, I recommend that the Commission urge Congress to develop some kind of reinsurance program to take up where the commercial companies leave off. Understandably, a commercial insurance company could not offer extended care insurance at an affordable premium because the risk is so great. There are several precedents already for government reinsurance: one would be the Federal Deposit Insurance Corporation which insures savings accounts now up to \$40,000 per account. This is an area that the commercial insurance carriers couldn't afford to venture into, so the Federal Government ventured into it. Another is the Federal Flood Insurance Program. Commercial insurance carriers could not afford the very great risk of providing flood insurance, so the Federal Government has ventured into that. A third area is that of theft and burglary insurance in very high-risk areas, which commercial insurance companies feel that they cannot afford to enter. The Federal Government has entered that.

I cite these three precedents of Federal Government reinsurance of otherwise uninsurable risks as precedents which could be used by the Commission in recommending to Congress some federal program of reinsurance for custodial care. In addition to that, as I mentioned earlier, I suggest strongly that Medicare be amended to provide some long-term custodial care.

The next area I would move to is the possibility of opening existing government facilities to HD patients. For instance, Dr. Arthur Davis, a psychiatrist, is doing HD research at the Veterans Administration in Palo Alto, California. He cannot find a sufficient number of HD patients. This is his desperate need. The facilities are there, and although I'm a veteran myself, I don't think there's anything sacred about the word "veteran." I think these government facilities should be available to HD patients, possibly to other neurologically distressed persons. At the same time, these patients would provide people like Dr. Davis with the patients who are needed for care. The VA and Public Health Service hospitals, perhaps, could be combined care/research facilities.

My final suggestion is something that I came across rather recently which I think is really quite exciting. It is that HUD (Housing and Urban Development) is rather regularly repossessing convalescent hospitals which have proven to be financially unfeasible. They can't make it. Concord, California, which is about 25 miles east of Berkeley, has been very overbuilt, and a number of convalescent hospitals are going to be repossessed and auctioned off to the highest bidder. One will be auctioned off next month. A convalescent hospital--a gorgeous thing--called RSVP was auctioned off last month in Carmel. RSVP is an acronym for Rehabilitation Service Volunteers of the Peninsula. This beautiful facility was built for neurologically handicapped people and has some unique features which would lend themselves to HD patients. It's gone now, but there will be others. I would leave this with the Commission just as a sample. These sort of things are coming up.

One of the doctors who testified earlier--the lady doctor, Dr. Moeller--suggested, as I recall, that there should be such facilities near the homes of people; in other words, there might be many of them. I don't know if that's practical or not. If a few of these places (three, four, or five) around the country could

be purchased, they could provide regional care facilities for HD patients, possibly for other neurologically handicapped people, too, because it might be difficult to get funding just for HD patients. These could be combined research/care facilities, particularly if they were located near medical centers. My specific suggestion here is that the Commission ask Congress to make these facilities available first (and on a preferred basis) for HD patients; that the Congress should make it possible that federal grants be made available to purchase the facilities, staff them, and operate them.

GUTHRIE: I'm very interested in your comments on the insurance. If you would, perhaps Nancy might be allowed to pick your brains a little further for the Commission. I think we might like more documentation. Right, Nancy?

N. WEXLER: Right, particularly if you know of any insurance for domestic care or homemaker chores. It seems almost impossible to get any insurance to cover that.

COMMERFORD: From the point of view of a commercial insurance company, it's just not an insurable risk. Insurance companies are insuring against possibilities, not probabilities. In other words, if a family that had a history of HD came to them and said, "We know we have a situation here where we're going to need home care. Can't you offer us some insurance to pay that?" that's like buying fire insurance for a burning building.

N. WEXLER: You can get paid for skilled care, but you can't get paid for unskilled care. Is that correct?

COMMERFORD: Some policies do provide for some limited home care; but, again, it's for a rather short time.

VOICE: Will you have a comment from the floor? We have the experience of my wife, Mildred, being home. My company had the foresight about five years ago to get major medical. They got it through Travelers Insurance. We were just a small company and had to have a minimum of ten people in the insurance setup, which we did get. About five, six, or seven years later we began to draw on that. They are actually honoring our claims for a nurse in the home, and they've been honoring this for over a year at about 80 percent of

the cost. Unfortunately, the premiums are pretty high, about \$150 a month. They have about doubled from what they were a year ago, but we're getting very appreciable benefits. This is one that is working out, fortunately, but it is expensive.

COMMERFORD: If you read the policy there is probably some limitation on the number of days that this coverage will be provided.

VOICE: Yes. I think most policies had it, but Travelers did not have that much of a limitation. I think it goes on for a number of years, actually. It looks good at this point.

CUNNINGHAM: Could I make just a very brief point on that? My wife has HD. We had a nurse for approximately two years because our youngsters were from 13 down to four or so. Our insurance policy paid for it to the tune of \$280 a week for 45 hours for this registered nurse. She helped my wife a great deal emotionally, and what have you. Finally, to conclude my comments, our major medical was \$50,000 per person. With 50 days in a psychiatric hospital and the nurse, it exhausted \$32,000 of our major medical, so we had to drop the nurse. She's carrying on, is quite active and doing the wash, and what have you. I was with another company, which I left seven years ago not anticipating this, that had \$365,000 worth of major medical coverage. You can use it up awfully fast, is my point.

M. WEXLER: The time limitations are very great, so just one minute. Yes, sir.

VOICE: A question in reference to life insurance for this gentleman on the panel. Could you comment on life insurance policies for HD patients or at-risk patients? My question is based on the fact that I renewed and updated and upgraded my insurance program last year to my satisfaction, probably, for the long term. I was interested in insuring my spouse. One company, of course, was very willing to insure me for a very large policy (for a very large premium, of course). I was interested in that because I want to take care of my insurance program now and not worry about it. But they would not insure my spouse, of course, because we were honest, and we didn't insure with them. This is a problem.

COMMERFORD: Very quickly, a person who had purchased a life insurance policy not knowing that he or she was at risk would not

have committed fraud. If they discovered subsequently that they were at risk, the insurance company couldn't cancel the coverage. All policies have a two-year clause whereby if a person conceals something of a material nature, that's fraud. The insurance company could contest a claim or they could refund the premiums. Beyond two years, even if one had concealed it, they couldn't refuse the claim. People who manifest HD and who already have insurance have no problem as far as life insurance is concerned.

The second category is that an existing HD patient couldn't buy life insurance for the same reason you couldn't fire insurance on a burning building. The third thing is that I called the Medical Director of the New England Mutual Life Insurance Company in Boston, and talked with him at some length about this because I've never had occasion to insure an at-risk person. He checked the records and called me back and said that in seven or eight years they'd only insured one person, and the premiums were just about double the normal premiums for an at-risk person.

M. WEXLER: I want to thank the witnesses. I appreciate their contributions. I'm going to call on the last group before we have dinner. Dr. David Comings, Dr. Al Giordano, Mr. Gene Bartolotti, and Mrs. Gayle Soergel. Dr. Comings, would you lead off, please, and give us your title, institution, et cetera.

TESTIMONY OF
DAVID E. COMINGS, M.D.
DIRECTOR
DEPARTMENT OF MEDICAL GENETICS
CITY OF HOPE NATIONAL MEDICAL CENTER

COMINGS: My name is David Comings. I'm Director of the Department of Medical Genetics at the City of Hope Medical Center. I simply wanted to discuss a genetics clinic we have which, in large part, takes care of patients with Huntington's chorea. I went through some of our records and wrote down some things about this, so I'll have to read this part of it.

A clinic is available at the City of Hope National Medical Center for the care, treatment, and counseling of patients with Huntington's disease and their families. The patients come by self-referral, referral by physicians, or referral by local Huntington's

chorea chapters. Although we serve primarily the Los Angeles area, some patients come from Northern California, Nevada, and Utah. If the patients have insurance, that is charged; if they have no insurance, there is no charge. About 20 patients are seen two to six times a year, and about 60 patients have been seen one to four times in the past four years. Of the latter, approximately 30 percent represent individuals who are at risk and wish genetic counseling and information about whether presymptomatic tests are available. Another 30 percent represent patients who are being cared for elsewhere but know of our clinical and research interest in Huntington's disease and come to the clinic to find if there are any new advances in therapy or presymptomatic detection. The other 40 percent represent patients that are on no therapy or on therapy which does not require constant monitoring. They come in about once or twice a year.

The patients who are seen up to six times a year are those who are being carefully monitored to maintain optimum effect from therapy or who have emotional problems and require supportive therapy. The most frequently used drugs are haloperidol, Valium, and Prolixin. In none of our cases has therapy provided a long-term total eradication of chorea. It generally provides 20 to 60 percent alleviation of the movement disorder. Early in the disease the benefits of a moderate decrease in chorea must be carefully titrated against the sedative effect of the drugs. In advanced institutionalized cases, large doses of medication are frequently necessary to control excessive movements.

Of the patients seen, all but three are ambulatory, live at home, and are cared for by spouses or family. These three are in nursing homes and are brought into the clinic periodically.

We've noted that the response of the family to an individual with Huntington's disease varies greatly. In some cases, the disease is taken in reasonable stride, with family members pitching in to help as much as possible. (These are the ones that join Huntington's chorea chapters and the ones that are visible here tonight.) In other cases, which we do not often see in these chapters, there has been total disintegration of the family with separation of the spouse and children to another household. These are some real tragedies.

About the research aspects of our institution, a number of individuals at the institution are interested in basic research into Huntington's chorea. In 1975 Drs. Goetz and Roberts and I observed that fibroblasts from individuals with Huntington's disease--these are skin cells that we take from the skin and grow in tissue culture--grew to a greater density in tissue culture than did normal cells. Drs. Barklay and Menkes, at UCLA, observed the same thing, and an observer in England observed the same thing. We feel this is an important clue, suggesting the disease may be a generalized membrane disorder affecting all the cells of the body. One advantage of this, then, is that we can do some studies on cells of a patient in the laboratory and the patients don't have to keep coming in and giving blood and samples.

Drs. Goetz, Roberts, and Laura Geary, in our Division of Neurosciences, are continuing to follow this lead by examining the conditions under which fibroblast growth may be enhanced, and under which we can make this difference between normal and Huntington's cells even greater and try to understand what the basic biological reason is for this difference in growth. We in the Department of Medical Genetics are attempting to find the mutant protein in Huntington's disease that Dr. Markham talked about. We are using what has proven to be a very powerful technique of separating proteins, and we have reasonable suspicion that the substance which is affected in Huntington's disease is a protein. This technique is to separate these things in two different dimensions in an electrical field. This allows us to look at up to 500 different proteins in a given sample, and there are certain characteristics we would expect of this mutant protein. We are using the brain bank as a source of this tissue and systematically going through different fractions of brains, looking for the mutant protein in Huntington's disease. We haven't found it yet. Individuals attending our clinic have been very cooperative in volunteering blood and skin samples for our research activities. Thank you.

M. WEXLER: Do you want to ask a question?

N. WEXLER: Quick. David, what is the proportion of your HD practice, would you say, in which the family is seriously disorganized by the disease?

COMINGS: I hesitate to say that from my own point of view, because it's just what it appears to be to me. It may be more disorganized than I realize, but I would say 10 to 20 percent.

M. WEXLER: I want to ask a question which will sound simple-minded, but I think it's of profound importance and I would like it for the record. Can there be a cure for this disease?

COMINGS: No.

M. WEXLER: You're saying no?

COMINGS: No.

M. WEXLER: Tell me why.

COMINGS: There is no cure for any genetic disease. By "cure," I mean the eradication of the genetic defect so that the disease no longer exists. Like diabetes, you don't cure diabetes, you treat diabetes so that you have no problem with it. But diabetes never goes away. We may find a way to more effectively treat Huntington's chorea so that the symptoms are not severe, or so the patient can live a totally normal life. Genetically speaking, it's impossible to cure Huntington's disease.

M. WEXLER: Unless there were a means of some genetic engineering which would alter the gene, you would have no cure.

COMINGS: Even that wouldn't work, Milton, because you'd have to go back to the original conceptus to change it then, and you could not.

M. WEXLER: Can there be interdiction of this disease so that the symptoms don't appear?

COMINGS: Yes.

M. WEXLER: If this Commission were to make a recommendation about a series of strategies and structures--take a tissue bank as a structure and a workshop as a strategy--that would be applicable to Huntington's disease, do you see it as possible, as feasible, as useful that such a series of strategies and structures for

Huntington's disease would also be applicable to other neurological diseases, degenerative diseases, and hereditary diseases?

COMINGS: Absolutely. For example, this technique that I talked about is applicable to every genetic disease that there is in existence, especially in both dominant and recessive. Recessive diseases are almost invariably an enzyme defect where it's more effective to try to figure out from what's wrong with the patient what metabolic pathway is involved and then determine the likely enzymes. We can't do that in Huntington's because we have no idea what metabolic pathway is involved. A kind of a shotgun approach like the one I'm talking about (to first identify where the mutant protein is, what its distribution is, purify it, characterize it, and find out what kind of protein it is) would then allow us to provide more rational therapy. It would also, I think very importantly, lead the way to possible presymptomatic detection and detection by amniocentesis.

M. WEXLER: Do you see any advances in research and research findings connected with Huntington's that might redound to the benefit of other dominant gene diseases?

COMINGS: Absolutely. I think it's the same rationale for developing a brain bank, for example. I came up in the elevator with Dr. Katz, I think his name was. He said his interest was in the autosomal dominant cerebellar ataxias. I said, "Do you have any brain material?" He said, "No." That's the first step. The same rationale and approach could be applied for that.

M. WEXLER: Is Dr. Barklay here?

BARKLAY: Yes.

M. WEXLER: David, did you want to say something?

BARKLAY: Just one comment from the floor. There's a very brilliant statement of Isaac Asimov's, about ten years ago, which was:

"If you ask any famous scientist whether something is possible and he says yes, he's probably right; but if he says no, he's probably wrong."

BARKLAY: [Laughter.] I do feel that genetic engineering techniques go so far as to insert new genetic material into people after conception; and that if Huntington's disease is the absence of a gene product, a cure, in the real sense of the word, is possible. I don't think it's beyond the realm of possibility, if Huntington's disease is the insertion of material which should not be there. But, for all practical purposes, it could be excised or switched off so totally and completely that an effective cure would be possible.

COMINGS: Let me defend myself [laughter].

M. WEXLER: I wanted to get the Asimov quotation in. Dr. Barklay has said that "If a scientist says that it is possible, he's probably right; and if he says it's impossible, he's probably wrong."

BARKLAY: That is correct.

COMINGS: Let me modify what I said before. I think in certain recessive diseases it's going to be very possible to effect a cure by genetic engineering. The distinction there is that if an enzyme is being affected, you could take cells out of a patient, insert the proper genes, put those cells back into a patient, and then the blood circulating through that organ would then have the metabolic defect cleared. However, if the gene which is affected is synthesizing a protein which is used in the immediate vicinity (say in the caudate nucleus of the brain), then I would like to have Dr. Barklay tell me how he's going to approach that.

N. WEXLER: We'll fund you both to try it [laughter].

M. WEXLER: We'll get Dr. Barklay to submit for a grant and see that he gets funded. Who is next in our group? How about Gene Bartolotti.

TESTIMONY OF
GENE BARTOLOTTI
SOUTHERN CALIFORNIA CHAPTER
COMMITTEE TO COMBAT HUNTINGTON'S DISEASE

BARTOLOTTI: My name is Gene Bartolotti, and I'm 63 years old. I am one of seven children in our family. Apparently, I have

escaped HD so far. Two of my brothers and one sister have died from the disease, and I have one younger brother that's in a convalescent home now.

My mother and father immigrated from Italy and landed in Oklahoma; I'll never know why [laughter]. She was ill for years. She contracted the disease, which we did not know, in 1929. She died in 1944. In the meantime, I had come out to California in the middle of the thirties (in the middle of the Depression, too). We had brought mother out to see if we could get some help for her from the County Hospital, and also from our private doctors. Of course, it was not diagnosed at that time.

We knew nothing of Huntington's disease until about 1960, when I had an older brother that was admitted to Camarillo State Mental Hospital for a mental disorder, and he was there for many years until he passed away. We were called there in 1960. Some doctor--I don't remember his name--did give us the first indication that it was Huntington's disease. Of course, after that it was the brother that spent seven or eight years in the Veterans' Hospital. Then I had a sister that--she had a very good husband that took care of her privately. He spent a lot of money to find a cure for her, which he never did, of course. Now I have the younger brother that's in a convalescent home.

These brothers and sister of mine all had children. I now have a niece that has just turned to the age, and now she has contracted the disease. She is 44. This will go on and on. What I'm trying to point out is the fact that if you take all of these ancestors and take half of them, 50 percent, that may contract the disease, and you put them in an institution, it costs thousands and thousands of dollars to keep them anyway. This is why I think the Commission should be listening and building special facilities for these people. Let these wonderful scientists and doctors have the money that they need for the work that has to be done, along with us clipping coupons, trying to raise money, garage sales, and what have you. This is all I wanted to say. Thank you.

M. WEXLER: Thank you very much. Marjorie, do you want to ask a question?

GUTHRIE: Gene, just for the record, could you possibly make a statement about how you feel not having gotten it and seeing other

members of your family being affected? Has it affected your life in any special way that you might like to say?

BARTOLOTTI: What it's done for me, I have three children, six grandchildren, and the more material I read the more I wonder whether they're free or not free of the disease. I've always taken the attitude that for some reason I didn't get it. That's why I want to work towards those that do have it. The other thing is that in each one of our families there has been a family dislocation. As far as the children of the younger brother that I have now in the hospital, it's a tragedy. They're coming around to see him once in a while. Thank you again.

M. WEXLER: Dr. Giordano.

TESTIMONY OF
ALBERT G. GIORDANO, PH.D.
CARMEL, CALIFORNIA

GIORDANO: My name is Dr. Al Giordano, from Carmel, California. I'm not an M.D., I'm an educator, an author, the father of four lovely children, and husband of a dear wife with Huntington's for 11 years.

I heard Dr. David Comings eight years ago, who I'm glad to see here again. My fellow Italian from Italy. I would like to say that even with a flat tire Bill and I got here in our little Porsche, alive. I'm 52, but I don't know how many years the good Lord has given me. If you know, please tell me.

I really feel that we have a tremendous resource of human resources. I'd like to indicate that I've traveled in the various states where I have taught and visited. Maybe I'm a little prejudiced, but I find that there is no shortage of intelligence among HD families. Therefore, I challenge this Commission: Let's save these dear souls for the contributions that they're going to be able to make.

I had a number of problems. Let me go back to my own personal life. Let me speak as a husband and as a father for some of you who are at risk and are trying to decide if you would have children. Going through the same situation, yes, I would. I might have two

or three instead of four. But if I have any sanity today, people, it's because of my four children. As most of us know, the early manifestations of HD start with personality changes. I had a lovely, intelligent wife that hardly knew the word "damn." All of a sudden I was an "s.o.b." I couldn't figure it out. We went through trans-actional analysis, psychiatrists. Thank God for our Catholic faith which I think pulled us through. I think one has to learn to live with the personality changes.

I haven't made a study of how many people desert their wives or are divorced a second and third time, but I am sure that we can be rather proud of HD families because I see that many people, a lot more have lived with the situation than have shirked their responsibilities, I would think. I think this is another important factor. Obviously, there are emotional problems, and these develop as the children grow up. They learn that their mother or father, the spouse who has HD, is somewhat different. It certainly affects their social lives. But if they have enough faith in their parents, I think that they will make it.

I'd like to refer to a note that I thought of on the way down here. If I were asked what I can attribute my success to, I would have to say some of the opportunities made available to me by so many people, people who I've long forgotten. This is one of my real roles as an educator, that I feel maybe I'm doing just a little bit of good among the harm that I also do. I'd like to say that I'm firmly convinced that HD builds integrity, responsibility and, very definitely, character. In order to make this trip, my 13-year-old and 15-year-old are in charge of the house alone. They are 21 in maturity. I think Bill testified to that. My two boys are away at college. My wife, at the moment, is at the Pacific Grove Convalescent Hospital.

May I share with you a bill for \$4,400, Nancy? Just a brief comment about extended care, and so forth. About seven years ago I had to make a professional decision for my own career. Life got so hectic when my wife had four or five accidents. We lived on a narrow road. We had to sell our lovely home and move in town. At that time I felt, "Well, this might be a good time for a change." I was offered a full professorship in charge of the Business Teacher Program at the University of New Mexico, in Albuquerque. It wasn't until our doctor at the time, a neurologist from Stanford,

the Palo Alto Medical Clinic, Dr. Paul L. [Truche], said, "Yes, I know Dr. Arnold Greenhouse, the Chairman of the Department of Neurology," that I accepted the challenge and the new position. One of my fringe benefits was that Jackie had excellent care, basically free, at the university. Her condition worsened, and this is now about four and a half, five years ago. I kept my ties to the college where I was because I had superior, I thought, fringe benefits, and I had a two-year leave of absence so (post, salary, fringe benefits, et cetera) I had to return to California.

Upon our arrival, and after a stay in the hospital, my wife was placed in Ave Maria Convalescent Hospital, in Monterey, run by the Franciscan sisters. At that time it was \$525; today it's 800-plus dollars, and the average in California is well over \$1,000 for most extended care facilities.

I believe the challenge to the Commission and to the government, our legislators, is to do for us, the people, those things that we cannot do for ourselves. I am not a pauper, but because I make a decent salary and have some royalty income, my children do not qualify for any kind of financial aid. On top of that, Blue Cross did cover, under their obligation, 14 months at Ave Maria; and then they reneged and said, "Well, we're studying the case." After eight months they said, "We feel that she is just getting custodial care," so I paid a \$6,000 bill.

We brought her home, and I want to say this. Because I felt at that time that without any relatives or other support it was essential that their dear mother, who was no longer an asset but a liability to the family, had to be removed, because I felt that, unfortunately, she would be doing more harm than good to the children. It's pretty tough to put it in my pipe and smoke when I have to tell my wife, "You're wrong, dear. The children are correct." Some of you may have had to live with that kind of a decision-making situation.

Anyhow, she did come back home for about a year and a half, two years. Over the years we've gone through all kinds of domestic help. This is another burden that we share. Even though we can't afford domestic help, if you've got four toddlers--my youngest, Maria, is now 13. She does not remember ever having had a well mother, but she's done much over the years to care for her mother. How do you measure those kinds of problems. I do think, thank God,

my children are stronger and better for it. There are no drugs; there's no alcohol; none of them smoke. Maybe these are my rewards. As Marjorie has said so often, it's not so important the number of years you live, but the quality of life. I thank God for having had my four children as long as I have. I don't know that I'll go home and find them there, or that I'll make it home, so let's be honest and practical about the situation.

I think the extended care question that Bill shared with you is one that I'd like to reinforce and state as a strong need, not so much for the care (which is also very important), but I think as a resource area where you have together 20, or 50, or 100 patients with HD that can be given better treatment, better care, better love, and hopefully find a cure that is so essential. Thank you.

M. WEXLER: Thank you very much. Mrs. Soergel.

TESTIMONY OF
GAYLE SOERGEL
YUMA, ARIZONA

SOERGEL: Thank you. I'm Gayle Soergel, from Yuma, Arizona. I'm here to speak about our family and how we faced HD. My husband, too, was a history teacher, a coach, and a professional athlete. He died a year ago last December. Yuma is a rather small city, but I would like you all to know that there's probably no one in Yuma who doesn't know about my husband and love him very dearly. He contributed a great deal, and our city, our Country lost something special when he died.

I think it's very important that those persons in the medical profession know all the facts and know them accurately before they discuss anything with HD families. We were given so much misinformation. One example is that my husband donated brain tissue. I had to go to the hospital at the last minute to sign some papers following his death. (He had been dead about 45 minutes to an hour.) The pathologist sat there and asked me when my husband had the flu. I said, "What do you mean?" He said, "The flu that brought on the HD." Not to mention a young neurologist at the VA Hospital, in Tucson, who told me that my husband and I were very unfortunate that we had sons; if we had daughters, they wouldn't be at risk. I didn't argue.

Nursing care is very expensive. We paid close to \$700 a month. Thank God we had the money, the resources, through the VA, through Social Security, and through a disability program at Yuma High School, where my husband was employed. All of the teachers and the custodial personnel, et cetera, signed up. We had to have 100 people sign up to get the program, and they signed up because of my husband. I might also add that my husband put me through school while he had HD so that I could take care of the family after he was unable to do so.

I think it's important that when someone you love is in a nursing home you live near them, because there is nothing more hellish in this world than being 100, 200, 300 miles from the person you love because they're in a nursing home. I think it is also important that those persons who work in the nursing home teach you how to care for your loved one. Nothing gave me greater pleasure than to go to that nursing home every day and feed my husband, aspirate him, do anything I could to take care of him, because I loved him, and I wanted him to know it.

I think it is important for the media to be absolutely saturated with any and all information that we can give them about HD. Last summer, in an effort to forget some of the pain that I had, I traveled throughout the United Kingdom. There was nowhere I went, there was no one I talked to who didn't know about Huntington's disease because of a very fine documentary broadcast put together by the BBC. I was thrilled to death, and I would like to see that same thing happen here in the United States.

I would also like to say to all HD families don't be afraid and don't be ashamed. Say anything and everything you can about HD. Let the world know about it.

I want to also say that my husband underwent neurosurgery in 1973. It was a disaster. If you are contemplating it for your loved one, think very seriously before you go ahead and have it done.

As far as brain tissue being donated, I think this is extremely important. It was my husband's last wish that some way, somehow, he could help his sons so that perhaps they wouldn't have to face what he has faced.

I want to say that I've seen and heard some very beautiful people here today, and I want to thank everyone.

M. WEXLER: Thank you very much.

N. WEXLER: I'd just like to ask one question of parents who have children at risk. There's some question about whether or not a predictive measure should be developed. We often hear from scientists particularly as to whether this measure should be encouraged. I'm just curious: as parents, what are your feelings about this?

SOERGEL: I have a very definite feeling about it. If you can find a cure for Huntington's disease, and you want to find out whether my children will have it, fine. If you have no cure, I don't want them to know.

GIORDANO: As you know, I have four children, and I think they each had different parents, because they're all so different. Therefore, they're different individuals. I'd have to say that I'd have to leave the answer to your question to each of them. I would say that perhaps one of them could handle the answer both intelligently as well as more emotionally; another may not. I would have to say that at a certain age they will make their own decision. I'd have to say even age 21, even though I feel my children are very mature at a younger age. I do not want that responsibility, as a parent. I think I would make that decision as I would about their education, or their religion, or their safety. I think it becomes a very personal decision that each one of them must make. A lot of people have a more disastrous illness, whether it's cancer, or whatever. There's always the question, "When do we tell the person?" or "Do we tell them?"

M. WEXLER: Thank you very much. I think we have to break at this point for dinner. Let's reassemble here in exactly an hour, which will be 7:35. Thank you.

[Recess.]

E V E N I N G S E S S I O N

M. WEXLER: Why don't we begin right now. The first group that I would like to call: Ruth Krueger, Mrs. Lyndon Jerry, Mary Boyle, and Mrs. John D. Little. Would all of those people come up, please. April Morgan? April, would you join this group? I wonder if we could hear first from Ruth Krueger.

TESTIMONY OF
RUTH L. KRUEGER
TORRANCE, CALIFORNIA

KRUEGER: My name is Ruth Krueger. My address is 2550 Pacific Coast Highway, Torrance, California. I have had Huntington's disease approximately 15 years. I am now 48.

I have had a Social Security card since 1937. I have worked off and on since then. The balance in my Social Security account is \$28,319.36. I am now totally disabled and find it quite difficult financially. I have been refused Social Security. I was refused Social Security because I lack \$50, or one point.

I knew I had HD but refused to admit it. Therefore, I wouldn't go to a doctor until 1965. He proved my suspicions. However, I was stubborn and went back to work in 1970 for nine months as a teller. I fell all of the time at work, dropped coins, et cetera. I then filed for Social Security. During that time, I was refused three times, and told I would have to wait until I am 63. I told the girl I would be dead by then, according to statistics. She had the nerve to tell me very coldly I should apply for welfare. Naturally, I was in tears.

I wrote to many senators (Charles Wilson, Bob Finch, Al Bell, and Ted Kennedy) who in turn referred my letters to James B. Cardwell, Commissioner of Social Security. I had a final court hearing October 18 of 1974 by a so-called unbiased judge. He was a judge for Social Security. He, his secretary, my husband, and myself--I didn't have a chance. Five years ago, my husband had a severe heart attack. Therefore, we had to sell our home and buy a mobile home. It hasn't been easy on us. I contacted my family attorney to attend the hearing. He was in court that day, so he sent a letter in my behalf to the judge. Still, it didn't help

my case. So far, I have contacted seven different attorneys, all of whom wanted enormous fees. I turned them down because I couldn't afford them. I found one thing for sure: We with HD need civil lawyers who won't charge us. I have all of the original papers on my case, if needed.

My mother had HD and was in a mental hospital for 15 years before she died. If we had a rest home for my family (three now with HD) our prayers would be answered. I would like very much to counsel anyone who has HD in their family. However, my finances will not permit me to help. I also had to give up driving seven years ago, so I have no transportation to help these people.

I would like to quote an article from the National Inquirer, which is of 9/76. It says:

"Crime does pay. Jailed rapist gets \$12,300 cash and \$340 a month for life from taxpayers. The Social Security Administration, in a disgusting mockery of American justice, has given a convicted rapist \$12,300 cash and is paying him \$340 a month while he sits out a 15-year jail sentence. If he is paroled he will have received far more than \$20,000 in benefits. What the Social Security bureaucrats don't know--won't admit--is that James Farley owes a great debt to his victims and society. Instead, his victims and society are paying him."

Thank you.

M. WEXLER: I find that a very interesting story. Could you tell me how old you are now?

KRUEGER: I'm 48.

M. WEXLER: When did you start working?

KRUEGER: When I was about 17 I started work.

M. WEXLER: You started working around 17, and you worked until when?

KRUEGER: Until '70, but I didn't work full time. Actually, I've worked most of my life.

M. WEXLER: Do you have any idea how much you contributed to Social Security?

KRUEGER: Yes, I do, over \$28,000.

M. WEXLER: How much did your husband contribute to Social Security?

KRUEGER: Quite a bit. I don't have his figure.

M. WEXLER: Would you say that it's at least the amount that you have contributed?

KRUEGER: More.

M. WEXLER: More?

KRUEGER: Much more, yes.

M. WEXLER: Between the two of you you've contributed probably well over \$50,000?

KRUEGER: Right.

M. WEXLER: And you are not entitled to any Social Security benefits whatsoever?

KRUEGER: Nothing.

M. WEXLER: Until you're 65?

KRUEGER: Right, until it's too late. Sixty-three, excuse me.

M. WEXLER: I find that really extraordinary. You've made such a very large cash contribution to a system that's supposed to protect you and preserve your health and rights. All you have done is pay in and there's no pay out, obviously.

KRUEGER: Really. It's a very sick situation.

M. WEXLER: You say that you cannot get legal representation without very substantial legal fees?

KRUEGER: Right. I have to have a civil attorney, which is higher. They are so much higher schooled, and they demand more money. As a matter of fact, one particular person quoted me \$5,000. If I'm going to fight for my money, I don't have that kind of money to spend.

N. WEXLER: You probably make too much money to have legal aid assistance, don't you?

KRUEGER: Yes; right. My husband is working, and I don't know what the whole situation is about. That really was a low blow when they told me to ask for welfare.

N. WEXLER: You probably couldn't get it. You probably make too much money to get it.

KRUEGER: Really. My husband was working then.

N. WEXLER: You were one credit off?

KRUEGER: Yes, \$50, one point.

N. WEXLER: Would you say that having HD might have affected your capacity to earn enough credits to be eligible for Social Security?

KRUEGER: Yes. I was not physically able to continue work, but I did go back for nine months. I knew I had it. I kept tripping and falling. They made another point, that I didn't go to the doctor soon enough to find out if I had HD. You know, you're not going to admit you have it until you really know. That's why I didn't get it, I guess.

M. WEXLER: I suppose the moral of that is that the only people who are protected in society are the very rich and very poor, and anybody in between is totally impoverished, really. They're desperate and without aid. Our system is designed to neglect that vast middle group which contributes so much to society.

KRUEGER: Right.

M. WEXLER: Thank you very much.

KRUEGER: Thank you.

N. WEXLER: Can we contact you for further documentation of your case?

KRUEGER: You sure can. I've got all my papers at home.

N. WEXLER: Thank you. Terrific. We'll be in touch.

M. WEXLER: Mary Boyle, please.

TESTIMONY OF
MARY GUTHRIE BOYLE
PRESIDENT
SOUTHERN CALIFORNIA CHAPTER
COMMITTEE TO COMBAT HUNTINGTON'S DISEASE
PRESENTED BY BETTY LITTLE

BOYLE: Good evening. I'm Mary Guthrie Boyle. I'm President of the Southern California Chapter of CCHD. Due to the fact that I'm having a big problem talking from a coughing situation, I have written my testimony, and Betty Little is going to read it for me so it won't be interrupted.

LITTLE: You can ask her questions afterwards. She can talk.

"I, Mary Guthrie Boyle, President of the Southern California Chapter of the Committee to Combat Huntington's Disease, would like to inform you of just a few of my problems with Huntington's disease. I was married to Woody Guthrie, and since he had HD our children were, of course, at risk. My girls grew up with the terrible fear of getting the disease that took their father's life. Having seen Woody in the later stages made it more horrible to know that this was the way they, too, might be one day. Our son, Bill, was killed at the age of 23. I look back now and I realize that probably it was a blessing. At least he won't have to end up like his sisters.

"Our oldest daughter was only 30 when she entered the hospital. She had a nervous breakdown which we felt sure was the beginning stages of Huntington's. She attempted suicide and was a very

miserable, mixed-up girl. She already had four lovely children whom she would never be able to care for again. We tried to make life easier for her, but mentally she could not cope with life. She knew something was wrong but didn't know what. It's a trait with HD patients that after a member of the family has the disease they have dreaded most of their life, they never really admit they have it themselves. She has been in a hospital now for eleven years and is fast becoming a vegetable. She weighs 83-1/2 pounds, can't talk, walk, and it's very difficult for her to swallow. In my view of thought she would be better off out of this life. Now her four children are playing the waiting game to see if one, all, or none of them go as their mother has.

"After our oldest daughter was in the hospital eight years, our younger daughter, Sue, was diagnosed HD. This is when we felt life much too cruel to live. Her life now is doomed and another grandchild at risk. She is very lonely and depressed. She avoids visiting her sister, knowing she will be in the same condition soon. She is still taking care of herself. Her walking is difficult. She falls down often and drops things badly. The involuntary movement is slight at this time. She has told me she will never enter a hospital but will take her life first. I personally do not blame her.

"Most HD patients do not like to socialize because of their problems with speech and walking. When they are in public, most people believe they are drunk. When eating, they drop food, silverware, et cetera.

"I know of no other disease that is as hopeless and helpless as Huntington's. Most at-risk families spend at least ten years worrying if they will get it. When the day finally arrives, when there is no doubt they have HD, they can expect their life to last about 12 to 15 years, and a good share of that time in some hospital.

"Most hospital employees have no knowledge of the disease, which makes caring for the patient unsatisfactory, to say the least. They assume that if the patient can't talk, they also don't understand. They don't include the patient in any activities, which makes their life more lonely. Now we come to the doctors' problems. Most doctors have very little information on Huntington's. Some

will accept literature if you can furnish some, and others don't want to be bothered. The result of this is most medication is on a trial basis--just a gamble. Physical therapy would be beneficial to the HD patient to keep them walking longer and retain some use of their arms. MediCal will not provide this because the disease is terminal. At the time we have only experimental medication except for tranquilizers. It's very difficult to find a convalescent hospital that will take an HD patient in the later stages because one person alone cannot take care of them. It takes two aides to feed, change their bed, et cetera. The hospital staff feels it's too costly to spend that much time with one patient.

"I know now that at my age I will spend a good share of my remaining life caring, hoping, praying and crying--helping take care of my girls. My only hope is research and at least a way to arrest or control this disease. After hearing the testimony of the doctors at this meeting, I do feel much more encouraged in this field and great hopes for my grandchildren and everyone else's. Thank you."

GUTHRIE: I just want to say that Mary was Woody's first wife; I am the second. I think that I love her dearly because she has been willing to share with me what is more important, not because we're the first or second wives. In relationship to Woody, we have found the most important common denominator, and that's to help others who are suffering with Huntington's disease. I truly love her, and I do try to help her. I am proud that she is the President of our Committee to Combat Huntington's Disease here in California. We do work together. I think this is a marvelous opportunity, and I wanted to say it to her.

BOYLE: Thank you, Marge.

M. WEXLER: We've been hearing several times today that kind of paradoxical thing, that tragedy has its special rewards, haven't we? I mean that it does something by way of cementing families, by way of creating some kind of caring attitudes. What I was remarkably impressed with was the sense of the expression that came from several people, that in the face of this kind of threat or this kind of illness time became really important. Most of us waste time a lot, don't we? When you begin to realize that time is limited and the ways in which tragedy or this kind of disease tells you time is

limited, it does make that time extraordinarily valuable. I'm glad that some people are really, really intent on making each day count. That's quite marvelous. Could we now hear from Mrs. Jerry.

TESTIMONY OF
LYNDON F. JERRY
PAST PRESIDENT
LOUISIANA CHAPTER
COMMITTEE TO COMBAT HUNTINGTON'S DISEASE

JERRY: My name is Lyndon Jerry. I'm the past President of the Louisiana Chapter, Committee to Combat Huntington's Disease. We keep hearing over and over the same tragedy for every family. My own husband was an attorney, and HD hit our family about 12, 13 years ago. His illness not only affected our family, it affected many of his clients. We actually became financially bankrupt. Because he was an attorney he caused other families to suffer great financial loss. But I don't think that's the important thing. I think the important thing is what it did to our family emotionally.

For five years, when it first started, my husband (who was a very calm, kind person) became very angry over little things. He began hitting the children, where he had never touched them before. Just kind of acted a little crazy. We went from doctor to doctor. Our family doctor told us that we had nothing to worry about, that there wasn't anything wrong with him. Then we went to another doctor, and that went on. We all ended up at a psychiatrist. The psychiatrist, after speaking with my husband, looked at me and said, "I suggest you come in to see me," and I became the patient. That went on until my husband actually displayed very obvious symptoms of Huntington's, and that's when we got the diagnosis.

If I understood correctly, Dr. Comings said that 10 to 20 percent of the families disintegrated. Being the President of the Louisiana Chapter, I was in contact with many of the families. I don't know how it is in other parts of the country, but in Louisiana I found that a good 80 percent of the families disintegrated. There were just very few that were able to take care of their patients. There were very few that were able financially to keep their patients at home. The only thing we had to rely upon was the State Hospital. The one thing I can say that the Louisiana Chapter did is that we did fight very hard for seven years to

upgrade our State Hospital to the point where we feel safer with our patients in the State Hospital than we would in any nursing home because of the checks that are put on.

Another thing that I think most of us have to live with--I know I do--I have my six children, 21 to 31, and five out of six tell me that they will commit suicide, not necessarily for a selfish reason (although they'd hate to go through what they have seen their father go through), but because they would not like to put their loved ones through what they have gone through and what they have seen an HD patient put their loved ones through.

I guess that about covers it. I think the Commission, hopefully, will make our legislators realize the amount of money it takes to care for HD patients and their families, because I feel strongly that the whole family is deeply affected. Perhaps at meetings like this you come across the more unique person, the person with probably a better financial situation, but I'm talking about the average HD family. I have found that because of the history of HD in the family very few--again, I can only talk for Louisiana--have any financial resources. Therefore, not only must the patient be taken care of--like our own family, when we ran out of money (paid thousands of dollars to the psychiatrist for all of the children, myself, my husband), then we had to rely upon the state to provide this. From a practical standpoint, if our legislators realize the amount of money that it does take, I think they will realize the practical benefit of research. That's all. Thank you.

M. WEXLER: I've heard it testified over and over again today that this is something of a bankrupting disease. Could you tell us the kinds of costs that you were exposed to that eventually led to your own personal financial bankruptcy?

JERRY: Yes. Because of the change of disposition my husband put off going to the office. I received phone calls from his clients, from his law partner, then even from the Internal Revenue. He tended to hide this. He was very shrewd. In fact, he kind of turned the tables around until he almost had me convinced that I was the ill one. It affected his business. Naturally, being an attorney, he could not competently handle the cases. Therefore, the income dropped gradually, like each year over a five-year period. Then going to the doctors cost an awful lot of money.

Like I said, we all ended up under psychiatric care, not knowing what we were dealing with. I thought that it was in my mind after a while. After having five competent doctors tell me that was nothing wrong with him, I had to assume it had to be me. It was like an Alfred Hitchcock movie, it really was. It started out, like we gave up the cruiser, we couldn't afford that; we gave up the help we had, we couldn't afford that; and I borrowed on insurance, and that went. We just reached a point where we lost everything.

M. WEXLER: It was basically the medical costs that were so very high?

JERRY: The combination of the medical costs and the lack of income due to the illness, yes.

M. WEXLER: Thank you. Nancy?

N. WEXLER: With six children, were you also trying to save something for them? I imagine that went also.

JERRY: Pardon?

N. WEXLER: I don't imagine you were able to save anything for your children?

JERRY: No, no, I wasn't. At first we kind of tore each other apart with our guilts, like when I reached a point where I had to put my husband into a hospital because I had two younger sons to support, and I hadn't ever worked. My children accused me of not caring about their father, and so on. We went through this hassle, I guess, for years. Now that they are grown adults, they realize. They seem to be handling it very well, I think.

M. WEXLER: What type of hospital did you put him in?

JERRY: The State Hospital.

M. WEXLER: That's all you could afford?

JERRY: Right. In fact, when I inquired at several nursing homes, they refused.

N. WEXLER: Why?

JERRY: Because they said that the patient had to be ambulatory.

N. WEXLER: Did you have a family history that you knew about, that the doctors knew about?

JERRY: No, we did not. I did check out the family history, and I found out that although it was misdiagnosed, out of the four children in my husband's family all four have had Huntington's disease.

N. WEXLER: It was never diagnosed correctly?

JERRY: No, it was not, until my husband.

M. WEXLER: Thank you. April, we've got you up there. I have you down for another group. Do you want to make a statement now?

TESTIMONY OF
APRIL MORGAN, R.N.
CLINICAL NURSE SPECIALIST IN NEUROLOGY

MORGAN: I'd like to introduce myself. My name is April Morgan. I'm pleased to say that through the Hereditary Disease Foundation I've been funded to act as a Clinical Nurse Specialist in Neurology, which means I'm a nurse merely for HD patients (and HD patients only).

By virtue of my position, I have seen just about all the ramifications of Huntington's disease that there are. The range of problems I could talk about would be genetic counseling, any kind of crisis intervention that may happen with a change in the family situation, a change in job status, a need for placement of a family member into a nursing home. I choose instead to talk about two primary issues that I think are very important for the Commission to stress: one being research.

We've heard a lot of talk about research. One thing that I try to do, which is one of my primary functions, is be the link of the family member with research. It's a big job. It's very difficult to share with family members, even though that's one of the greatest hopes, all the research that's going on to obtain tissue for

research. It's a very big job. I'd like to see more money allotted to this type of liaison of the families and of the patients and research. Enough on research.

Another very, very important aspect that I would like to stress is education. We've heard much on education for family members, much on education of physicians. I agree, the need is there. What I'd like to stress, however, is the education of the nursing staff. I, being a nurse myself from a very, very good nursing school with a baccalaureate program, never heard of Huntington's disease in school. I was not exposed to Huntington's disease until I had my very first patient on a psychiatric ward. I see this over and over again. People just haven't heard of it.

I often go into nursing homes; I see convalescent homes, the staffing in convalescent homes. The primary giver of care to patients in nursing homes are nurses' aides. They're not your R.N.'s, they're not even your L.P.N.'s; they are nurses' aides. They have the least amount of education. I go into nursing homes to say, "Draw blood on a patient," and a nurses' aide will come in. They don't talk to the patient. They talk over the patient, they talk around the patient to me. I resent this. So, good Lord, I wonder how the patient feels.

What I would recommend is a better education system for nursing homes, and this can be done. There are movements toward continuing education for nursing staff, but it's exceedingly inadequate. It can be done. I've offered to give inservice programs to nursing homes. They don't have the time. The problem is manifold, but I think it's one that needs to be addressed. I've heard recommendations for developing specialized nursing homes just for Huntington's disease. This is a good dream. To more appropriately address the issue, I think, would be to educate the staff members in nursing homes that already exist. I think that would be a goal that is started, but it just needs much, much more attention, and all of that would be for the better care of the patient.

M. WEXLER: I'm going to make a strange statement. You may be listening to the most important person in the world in connection with HD when you listen to April Morgan. I want to justify that statement. You see, there are a whole bunch of wonderful scientists on one side, and they have fantastic skills. One of the things that you may have listened to with some astonishment, because it came up

over and over again--I would like to emphasize it again--is that the kind of researcher or the kind of scientist who may have the answer for HD has never heard of HD. That means, in a sense, that there are a whole bunch of very basic scientists (with basic science knowledge) who have to be introduced to HD, who have to learn the clinical aspects of HD, who need an education in HD. As they develop their interest, they will need the kind of tissue on which they can make their investigations and find an answer for HD. That's at one end of that continuum.

On the other end of the continuum is the vast mass of patients, at-risk people, family members, who have the kind of tissue on which these scientists can work. In between, as a liaison, as a means of communication, as a means of data-gathering and tissue-gathering, there is April Morgan. Without that link, nothing exists. The scientists don't exist and the families are helpless and hopeless, without anything. I know I'm dramatizing her position, but I'm not exaggerating it. I'm really saying to you that we need more April Morgans, we need more scientists who are in touch with April Morgans, and we need more family people who are constantly using this kind of liaison to make their contribution to getting an answer. That's the reason our foundation was very intent on funding somebody like April Morgan to act as that liaison. We were very lucky to find her. I don't know whether we can find a series of April Morgans in this world, but we can approximate in some way.

I want to add to her recommendations that we make a very strong and urgent recommendation to Congress that we find more April Morgans. You know, there is a kind of biological research which is somewhat frowned on at times; nevertheless, it's called cloning. Maybe we can clone a few April Morgans (which means to produce exact duplicates), and we'd have a whole wonderful series of liaison people. I'm really talking to you people out there, that if any of you haven't talked to April Morgan, you are failing yourselves. No matter what, she's probably the most important person in the world you can talk to because you have an immense contribution to make to the solution to this problem, and she's the girl who can put you into the mainstream of what it takes to find an answer. We're going to find an answer. Thanks, April. Do you want to ask April some questions?

N. WEXLER: We just want you to teach us how to teach all the rest of the nurses. You don't have to do it tonight.

MORGAN: I've got plans.

M. WEXLER: We thank all of you for your testimony. We appreciate it.

LITTLE: I wrote a letter to the Commission. Won't that do?

M. WEXLER: Okay, do you want to just leave us the letter?

LITTLE: We're running short of time. Will my letter suffice?

M. WEXLER: Yes. Thank you. The next group: Mr. John B. Link, Deanna Link, Hannah Geer, and Sol Babitz. Mr. Link, would you begin, please.

TESTIMONY OF
JOHN B. LINK
VILLA PARK, CALIFORNIA

J. LINK: Thank you. My name is John Link. My wife's name is Val Link. We've been married for 35 years. We have three children, all in their twenties. My wife was diagnosed in 1969 as having Huntington's disease. At the present time she is under full-time care at our home. We've been extremely blessed with having a very wonderful person to take care of her, a very understanding person. For this, we are extremely thankful.

I didn't know quite what to say, but I thought that I would kind of briefly run through the situation in our family and describe the impact upon us and let it flow from there. My wife is an only child. Her father passed away when she was two years old. Her mother passed away about five years ago, with no signs of Huntington's disease in her family. We believe the carrier to be in the father's family, but it's been impossible to really track backwards in the family to confirm this.

I've known my wife since she was in early teens. The only indication that I might have had was a twitch to her left foot which was not controllable--it was infrequent--but which disappeared when she went to sleep, of course. As I look back now I can see that that was a symptom. In 1967, while we were in the Bay area, I noted that one of the corners of my wife's mouth twitched, and she could not really control it. Not knowing what it might mean, we went to

several doctors, spent a few thousand dollars and ran tests. Finally, the doctors agreed that it was something--I think they called it a tic, or something like that, but basically said that there was nothing that they could do about it.

We moved to Orange County in 1968. In 1969, when my wife was at Kaiser Hospital with one of our children, one of the doctors apparently observed this muscular movement and suggested she see one of the neurologists there. Very shortly, he diagnosed it as Huntington's disease and two neurologists later confirmed this. HD was something which was totally unknown to us, and we really did not know the impact of the diagnosis. I guess the impact of the diagnosis was devastating to us, to my wife, in particular, and it went through the various symptoms which the medical profession knows far better than I do. Basically, it had to do with complete rejection of the diagnosis, to the "Why me?" syndrome, and finally, "I'm going to lick this thing." Thanks be to God. In some way or another He gave us an inner strength to begin to live with this foe. We're very thankful that we have now come to grips with it and are able to live with it at the present time, and we hope that it continues this way.

I had hoped that one of our daughters, Deanna, would be here this evening. For some reason I don't see her yet. That's usually the problem with young folks, I guess. They have some difficulty in making engagements. I would say that I do know that it's constantly on their minds. It is constantly figured into their plans. We discuss the disease. We do not attempt to hide it. We bring it out in the open and discuss it. All of our children are college graduates; one, a lawyer. They've shown no indications to me yet of doing anything violent with their lives if they do get HD, and we're hopeful that this, of course, does not happen.

As far as my wife's treatment is concerned, from 1969 to 1975 she was under a neurologist's care in Orange County, Dr. Carl [Barth], who had treated only two HD patients in his some 15 years of service and had no ongoing history of treating them. In 1975 my wife came under the care of Dr. Comings, for which we are truly thankful. We feel that as far as medical help is concerned we've been most fortunate. When I talk with people and hear stories of some of the things and lack of medical attention, I can really have empathy with them and know that we really have been fortunate, if that's the right term to use.

I wanted to just briefly mention something about the problems that we have or have had. By the way, if we can get our daughter in, Dr. Wexler, she may want to testify, if it pleases the Commission.

M. WEXLER: She's here now?

J. LINK: Yes, sir. She just came in.

M. WEXLER: Come and join your father, as all daughters should do.

J. LINK: If I can just wind up what I would like to say, please, I would say that the problems that we have faced up to the present time have been rather minimal. I would say that one of our problems has been--and it's common to all of us, I believe--trying to find and afford adequate help to take care of our patients who have this disease. We advertise in newspapers and such as this, whatever means we can use, to try to find somebody. At the present time, as I mentioned, we're very fortunate to have someone in this capacity.

As far as finding affordable and adequate nursing homes for my wife, I investigated that. They started off at \$1,000 and better a month. We kind of ran from that in a hurry. Although I'm sure there are probably homes that treat this in Orange County, we're just not aware of them at the present time.

...certainly, had we known of the counseling that we now know about, we feel that we could have done a better job sooner in beginning to understand the impact of the diagnosis that was made. I would say this--and Dr. Wexler referred to it a few minutes ago--that in this disease we've found that our family has truly come together, that we have a common foe with which we are wrestling. If that is one of the good aspects of it, if that's the right way to say it, we feel that from this standpoint it has done things which might not have been done before. I think, if it pleases the Commission, that's about all I have to say.

M. WEXLER: Thank you. In that case, we'll ask Deanna Link to talk to us.

TESTIMONY OF
DEANNA LINK

D. LINK: I wish I could have heard everything my father said, because our viewpoints may be different. I don't know. I'm Deanna Link. I'm 27 years old, single, and I'm working in the field of social work. I'm a patient at risk, and my mother is currently in the advanced stages of Huntington's disease.

I'd like to talk to you briefly about some of the special problems this disease has caused my family and me. I recently took care of my mother for a period of two years, so I've had the opportunity to observe the manifestations of the disease closely. My mother is on the verge of being totally crippled. At this moment she is only able to walk just short distances. I can only carry on the simplest of conversations with her, as her rate of comprehension and retention is extremely low. My father has hired a woman who lives in our home and takes care of my mother full time. This is necessary because of my mother's inability to take personal care of herself and because of the existing hazard that she may accidentally hurt herself.

As I've observed the disease gradually debilitating my mother over the years, I've grown more and more afraid that this would also be my own fate. The possibility of losing my physical and mental abilities frightens me very much. A genetic counselor told me that the peak age of getting Huntington's disease is 35 years old. Even though I know it is impossible, I feel a tremendous amount of pressure to do all the things I want to do in my life before I reach that age. I'm planning to begin a master's program in social welfare in 1978. It is a two-year program, so I will be 30 years old by the time I finish. It doesn't give me much time to pursue the career goal I have of working to alleviate problems in poverty, and another goal I have of traveling extensively in foreign countries.

The disease has also posed another problem in terms of my decision as to whether or not to get married. When my mother first began to show symptoms of the disease, she was extremely nervous and easily angered. Over the years she has become more calm and lethargic. Her extreme change in behavior has put a tremendous amount of pressure on all the members of my family; in particular, my father. In spite of all the problems, my father has made a deep

commitment to his relationship with my mother. He has stood by her side through all the difficult and painful times. In so doing, however, he has made innumerable sacrifices along the way. Having seen what my father has gone through, I don't relish the thought of putting someone I love through the same arduous experience.

On the other hand, I must admit that I'm very afraid of being alone through the illness. Even if I were to get married, I certainly wouldn't expect someone to make the same commitment to me that my father's made to my mother before I were to actually get the disease. In any case, I have decided positively not to have my own children. Certainly, I want to avoid the possibility of my passing on this debilitating disease to others.

From an emotional standpoint, there is much more information that I could give you, but I find it difficult to express such deep feelings in this environment. I come before you as an at-risk patient, having briefly shared some personal feelings about how Huntington's disease is affecting my life. I hope my own testimony and that of other concerned individuals will move the Commission to react in a way that will truly help those that do not know and those that have already been stricken with this crippling disease. Thank you.

M. WEXLER: I think that everybody here appreciates the willingness to share that kind of experience and those anxieties. I would like to ask you questions which may be very difficult to answer. If you make the assumption that you don't want somebody to go through the same kind of torment and difficulty and problem that your father has gone through, and if you are simultaneously frank enough to confess your anxieties about facing that situation alone, what kind of structure, what kind of organization, what kind of program, what kind of what, would you feel might in some way represent some assurance to you that if by any chance this would happen to you, there would be some way in which you could be adequately taken of? Do you have any ideas on that subject?

D. LINK: I guess I'm not really certain about what you're asking me.

M. WEXLER: I'm asking, for instance, what kind of program could the government establish (what kind of program for social workers, for example, for nurses, for nursing homes, for counseling,

for psychological assistance, for genetic counseling, whatever) where you wouldn't feel desperately alone, where there would be some kind of services available to you in the off chance that you did develop the illness, where you would feel that at least there is some help and that you aren't really alone?

D. LINK: I really couldn't answer that. I don't have any proposal. I guess I haven't thought through that deeply about what could exist where I wouldn't feel the way I do.

N. WEXLER: Could I ask the question in a different way? If the disease didn't wipe out a family so totally financially, and if it wasn't so hard to find either good care at home or good institutional care, would you be as reluctant to get married if you really felt that there were other supports?

D. LINK: Perhaps not. That's really a difficult question, but I don't think so.

N. WEXLER: I would just like to say one thing which I've been responding to all afternoon and evening. It's small, but I consider myself an at-risk person, not an at-risk patient. To me, that makes a big difference.

D. LINK: I only refer to myself that way because that's the way I've been referred to.

N. WEXLER: Don't settle for it [laughter].

GUTHRIE: Along those same lines, Nancy, I wanted to comment. You said that at a certain age you might develop it; therefore, you're thinking that you may not be able to practice your profession. I would like to reassure you, because sitting right next to you is a man with Huntington's disease who has continued his profession, and he's going to speak about that. Unfortunately, you came here late today, and you have not seen the people who are functioning. You may have ten very good years in your profession long before you could not function in it.

D. LINK: I have very little contact with--in fact, I've had no contact with anybody who has the disease, other than my mother, and she could not.

GUTHRIE: That is why I'm so pleased that you're here and that you're going to hear the testimony of the gentleman next to you.

M. WEXLER: In that case, we'll call on Mr. Babitz.

TESTIMONY OF
SOL BABITZ
LOS ANGELES, CALIFORNIA

BABITZ: I'm Sol Babitz. I'm a violinist. My father died of Huntington's disease about six years ago. I became aware of symptoms around the same time. Previously, I thought he had Parkinson's disease. I became aware of having symptoms similar to his, the lack of control and motions, difficulty eating, sometimes speaking, and so on. I found that using a lot of vitamin B and other medication (tryptophan, and so on), and also eating at least 80 percent raw food, has enabled me to keep on functioning. I've noticed absolutely no slowing down in my reactions in playing the violin. I'm 65. I know that if I keep on the same diet and take a lot of the B vitamins, and so on, I'll just live a practically normal life from now on [applause].

M. WEXLER: I want to add, because I think he sort of modestly speaks of himself as a violinist--and he's an extraordinary violinist--that he's also an extraordinary composer. I'm sure that in all those areas he's functioning remarkably well, and we're very grateful that he's here. I hope that all the people sitting next to him and out there are really encouraged by that statement and that courage. Are there any questions?

N. WEXLER: No, I just wish I had a different dinner [laughter].

M. WEXLER: Raw food, vitamin B. I think it was Gene Roberts or Dave Comings who, just in passing, mentioned something about vitamin C and vitamin E with good reason. For several years our scientists have been speculating about vitamin C and vitamin E. I think many of you have heard me talk about it, because both of these are very effective anti-oxidants, and there is some reason to think that that might have some relevance to the ways in which the process proceeds in Huntington's patients.

VOICE: Excuse me, please. Was that vitamin E he said?

M. WEXLER: Vitamin E and vitamin C.

SIMON: He said vitamin B.

M. WEXLER: He's talking about vitamin B, yes.

BABITZ: B complex, in large quantities.

M. WEXLER: I was simply adding to his vitamin categories vitamin B complex, vitamin C and vitamin E. We had a very special reason for talking about C and E.

GUTHRIE: I think I've seen maybe more patients than most people in this room--most people, and on the whole. One of the things that I think is also important--and I observed it tonight--is that Mr. Babitz also has a loving wife and a concerned daughter who speak to him as a whole person, who treat him as a whole person, and I'm sure they've never done anything else but that. That's quite different from what a lot of people have done all across the country, because we didn't know any better. The fact is, we do know better; that where there is personal, tender loving care and understanding of what the course of this disease could be, we will see a different kind of patient than what we've seen.

M. WEXLER: I've got to make a speech about that, too. As I've said, my boss over here, who tells me what I'm supposed to do on the Commission, assigned me to the Behavioral group (that is, the group studying behavioral problems in Huntington's disease), and we've met in Bethesda to discuss behavioral problems and the kinds of things that have been talked about all day today. The reason I was put on that, of course, is that I'm a psychoanalyst, so I'm supposed to know something about behavior. Jennifer and I have been to Boston and listened to a lot of testimony, and over the course of years I've listened to a great many patients, talked to many families, et cetera. Today was remarkably impressive to me in terms of the psychological aspects of Huntington's disease. What has come home to me over and over again today is the enormous impact of loving care, of concern, of psychological interest, of the idea of the patient not being segregated off and separated from the family, of the deterioration which seems to follow from rejection and isolation.

In terms of all the people that I have seen, the patients who have testified, the families of the patients who have talked about

the patients, it's really dramatically impressive to me that given that kind of environment, the right psychological environment, it seems to have a remarkable impact on the progression of a disease which is mainly spoken of in neurophysiological terms. I guess the Commission would be thoroughly justified in making a very profound, a very important, a very significant recommendation to Congress and the President that these aspects have to be taken care of in terms of the way these patients are cared for by nurses, by doctors, in convalescent homes and elsewhere. We can look for medications and we can do all kinds of research, and all of that is of paramount importance, but in the process we must not forget that they are human beings. As Nancy said, they are persons, not patients. We'll hear from Hannah Geer.

TESTIMONY OF
HANNAH L. GEER
MISSION HILLS, CALIFORNIA

GEER: My name is Hannah Geer. What is the worst possible thing that could happen to someone you love? Death is the answer to most people. But death of what? Our earthly body as we know it? What about the loss of everything that a person was, personality, ability, ambition, motivation? That is a form of death, too.

I have been married to my husband for 28 years. For 20 years he was the man I married, until Huntington's disease. Since then he has changed until I hardly know him. Gone is his quick wit, the constant smile, the self-assured man that he was. In his place is an indecisive, docile human being. The only way you can remember how he was is snapshots, or friends relating past experiences, or fond memories. Because of HD this man has become a robot-type individual that goes through the business of living a day at a time. Any trace of his former self is gone from his family and friends as surely as if he were in the grave. Beginning senility at age 83 is a very sad thing indeed. Beginning senility at age 53 is a disaster. HD, the same as any brain disorder, is senility in all its forms. It is the worst possible form of disability because it strips you of your dignity.

It is so hard to conceive that this man, who was such an extreme extrovert, well-liked, with such a commanding personality, so ambitious, skilled in so many ways, should be reduced to this existence. I am indeed fortunate that he has a tranquil personality

and is not rebellious. He accepts his fate and does not complain about it. His wants are very few, and at this time I am able to handle them. It is the future that scares me to death. Unless HD is stopped soon, I know he will most surely go downhill. I have seen the effects of HD, and I know what is ahead.

People with HD need a place to go near their homes where professionals that are familiar with this sickness can take care of them. Many institutions are not staffed with people who do.

When I talk of dying personalities, I'm also speaking of my own. When you live with a spouse that's afflicted with HD long enough, you change, too. You become irritable as you try to cope with all the problems that come along. These problems range from trying to pay the bills to fixing a leaky faucet. Someone who cares for HD people need professional guidance, too. If you are in a moderate income bracket, this help is far beyond your reach. We need someone who can counsel us, so we don't lose our own identities.

Foremost in everyone's minds is a cure. That is all-important. But still, help us to cope with the stricken ones. I, along with everyone else in this room, am hoping that you will realize that we do have a terrible genetic disease here that does need special attention for it is as deadly as cancer, or heart trouble, or any disease imaginable. Even if it is too late for my husband, no offspring of his should have to endure this disease. We have one daughter and two beautiful grandsons. The thought that they would have to endure this is unbearable.

M. WEXLER: Thank you very much. Thank you all for coming and testifying. Could we have Jan Espinoza, John Starr, Iva Kline, and Molly Squirrel, come up, please. Jan Espinoza, will you start?

TESTIMONY OF
JAN L. ESPINOZA
VICE PRESIDENT
NATIONAL HUNTINGTON'S DISEASE ASSOCIATION

ESPINOZA: My name is Jan Espinoza. I am currently Vice President of the National Huntington's Disease Association and president of the local chapter. It's amazing how a person can live their whole life without ever hearing the words "Huntington's Disease." I never missed them, but from the moment I heard these

two words, I lost control of my future and my present, because the reality of HD in my family and the threat that looms over my head have changed the very person that I would have been. But it's the nature of this beast to fight anything that threatens, to hit it head on with all the intellect, talent, and enthusiasm available.

As soon as my family became aware of HD organizations, we began attending meetings and availing ourselves to the needs of researchers. I became President of the Family Membership Group of the California Chapter, Committee to Combat Huntington's Disease. Later, I was president of that chapter for almost three years, served on the National Board of Trustees, and was a member of the Executive Committee. Last year I was a founding member of the National Huntington's Disease Association, and I've already told you what else there.

I don't give you these qualifications for glory, but I do wish to demonstrate my intense interest, my long fight, and qualify my special kind of expertise. I have seen much, listened to many, to these families. These thoughts that I am about to give you reflect both my personal and their views. Many of the people have become close personal friends. In fact, one of the most painful things I face is the possibility of watching these friends, many of which are sitting at this table, as one by one we are taken by HD. Which ones of us will it be?

For so many years all of us have fought the personal and organizational battle of HD. We have fought the indifference and negativism internally and externally. We know this is the first time that anyone with the power to do anything about our problems has asked that all-important question: "All right, what do you want?" Eureka! That's what this day is all about. But, you know, it's really difficult to come up with those answers. Our problems are so multifaceted and so complex. We need research toward cure, control, and detection. We need involved, educated care; counseling; improved prolonged care situations. We need financial relief and education of the professional and lay public. Because of the longevity of the disease, the problems are overwhelming and ever-changing.

Of course, the Government can not and will not be able to solve all the problems for all of the people. One solution for a good many of the problems would be regional research, diagnostic and treatment centers. A family could visit such a center for a few

days, where a variety of help would be available. The patient could undergo a complete neurological and psychological evaluation and return home with a specific set of instructions with regard to medication and care to be carried out by his personal physician. In this way all patients could be receiving expert care. Further, during this time the patient could participate in existing research projects, a most worthwhile and necessary activity. At this time evaluation could also be made as to the need for physical, vocational, and speech therapy. The use of these available therapies is minimal and the possible benefits of increasing the active life of HD patients untested.

But, as we all know, HD does not affect one person singularly, but is a tragedy for the whole family. Radical changes in the spouses' lives are imminent and the ability to deal with them limited. Counseling could be available at such a center both to support and guide the spouse and to help the patient by his spouse's better understanding of how to provide a healthful home environment for him.

Then, of course, there are the children of the HD patient. What a burden they must endure. They (we) are called "at risk." But we are also children of HD patients, and we must make this distinction because even if the nature of HD was not hereditary, the difficulties that develop and the long-range changes that occur in the very person that this child would have become are major. Of course, the younger the children are when they experience this painful thing, the more devastating the effects. As the children age, a tremendous amount of pressure bears on them to make decisions about their future lives with regard to marriage, childbearing, and planning for the possibility of HD.

No one who has not lived in this den of fear and uncertainty can ever begin to understand it, families or friends, no matter how loving. Yes, there are no certainties in anyone's life. For the at-risk the uncertainties are just a little more certain. Finding a way to live with HD but not allowing oneself to be controlled by it is difficult. Counseling should be available to help the HD at-risk individual bring HD into perspective.

Beyond these types of direct aid to individuals, centers such as these would provide long-ranging changes in the total portrait of HD. Their existence would bring respectability and acceptability

to the disease and encourage dollars for research by bringing the disease to the attention of many. The problems HD persons face in the throes of an unkind and uneducated society would be altered by the notoriety the centers would provide. Once familiarity is established, so many other problems would be able to solve themselves.

Whatever the final recommendations are, it is imperative that we stress the importance of the effects on related diseases and problems. A center such as I have described would do work that could benefit many. This will give the suggestions acceptability, because no matter how devastating for us, HD does affect a small percentage of the population. If we attach ourselves to other genetic or neurological diseases, we do have a much greater chance of meeting our similar goals.

I continue my personal battle and have even geared my professional goals toward helping in our mutual battle. "Look to this day, for it is life." These are the words that I live by. But it isn't dying with Huntington's disease that I fear; it's living with it. Thank you.

M. WEXLER: Thank you, Jan.

GUTHRIE: Jan, one little quote. You used the words, "the proof was untested." I hope that your visit today and seeing all the patients who are here, like Mr. Babitz and some of the others, will tell you that there is proof that all of the things you said that could be done are being done; and when they're being done for the right people, are helping. There is proof. The question is to get it out to everybody.

ESPINOZA: Thank you, Mr. Babitz, because as of this day I had never known any patient that had sought out and really utilized these things that are available. I just have this tremendous feeling that if you ever took somebody and you gave them all the kinds of help you could give them, you could really make a difference in their life. My own father hasn't done this, and I'm ashamed. I really feel that these different things are there, and I don't think our patients are using them. I don't think the therapists are really fully aware of what the effects might be. I don't think that any of us have explored this area.

N. WEXLER: I just want to say that I'm glad you're going to be here after this day is over, because since I first met you three years ago--four years, five?

ESPINOZA: I've been at it for seven.

N. WEXLER: The amount of energy and enthusiasm makes me much more optimistic that you'll keep plugging even after this Commission is over.

ESPINOZA: Yes. Thank you.

M. WEXLER: Thank you, Jan. Mr. John Starr, please.

TESTIMONY OF
JOHN STARR
CHAPTER VICE PRESIDENT
NATIONAL HUNTINGTON'S DISEASE ASSOCIATION

STARR: Forgive me for reading my remarks. I feel extremely nervous up here. There's no reason I should, because I've been used to thinking in terms of all HD people as part of a family, which is perfectly natural, considering the nature of the disease. Still, it's difficult to come up and say anything that will have a significance for the Commission.

I am John Starr. I live in the Santa Barbara area. I am a technical writer, and I've been active in the affairs of HD family organizations for the past four years or so. At the moment I am privileged to serve as a chapter vice president of the National HD Association.

I would like to comment about my place of residence. I feel a special sense of mission tonight because I know a number of people who live in areas far enough removed from this site to make it extremely inconvenient (and, in fact, impossible) for them to be here tonight. I'm thinking particularly of one family that lives about 160 miles northwest of here, up the coast. I know they wanted very much to come, but the traffic, the condition of the patient in that family, the necessity of coping with two children who are in their early teens, I believe, make it impossible for them to be here. It's particularly for them that I'm speaking.

Of my own situation I'll say simply that about 15 years ago, when I was about 20, my sister and I first learned that our mother's violent temper, irrationality, and physical deterioration were associated with something called Huntington's disease, and that we were at risk. I am divorced and voluntarily childless. I'm particularly grateful to Deanna Link for her testimony. I'm sorry I wasn't able to hear earlier testimony. Deanna spoke, I think, far more eloquently than I would ever be able to bring myself to do of the feelings that haunt me and a great many other young, at-risk adults.

I'd like to second all preceding testimony--and that's a lot of it--which calls attention to three needs especially: (1) for increased research on HD and its treatment; (2) for more thorough dissemination of information about HD among health professionals and agencies (both private and public); and (3) for programs to help lift the crushing financial burden which HD imposes upon many families.

At the moment, however, I wish to address a fourth area of concern, one which has recently been touched on, and one which merits (and which I had been afraid might not receive) as much attention as the others. I'm referring to the need for personal and family counseling services aimed directly at HD. In my opinion, the social cost of HD, in terms of its psychological effects on non-patient family members (especially children), is immeasurable. No comprehensive program for the control of HD and its consequences can afford to ignore these effects. I have in mind a comprehensive program of personal and family counseling conducted through regional Huntington's disease centers and coupled with training programs and campaigns to increase public and professional awareness of HD.

The methods and goals of such a counseling program would have to be determined by the knowledgeable professionals overseeing it, of course, but I think they might aim at the following: First, the counseling of HD patients to give them a clear understanding of the disease, a realistic assessment of their limitations during its progress, and a firm foundation for self-respect in the face of circumstances beyond their control. Patients would, I hope, be helped to find useful activity, perhaps even some kind of involvement in this or some other HD program within the limits of their capacities. They should also be helped to comprehend the emotional

needs of their children especially and be trained in ways of minimizing the effects of their condition upon the other members of their families.

The second aim would be the counseling of adult, non-patient family members, especially husbands and wives, to help them understand the emotional problems of victims, and to help them find effective ways of coping with the attendant complex of problems that they face (medical, financial, emotional, et cetera) during the various phases of the disease. Such counseling is crucial, I think, at the time of diagnosis and throughout the period of onset. That doesn't mean that I think it should be done away with or neglected after that time, but particularly important at that time. Special counseling should also be offered for those facing the decision of whether or not to place a loved one under institutional care.

The third aim is special counseling for children, to help them understand the behavior and the requirements of an afflicted parent and to cope with the resulting emotional stress within the family. The emphasis of this counseling should change as the child grows older to help him or her deal with the at-risk status and the accompanying problems of facing an uncertain future.

I had heard the suggestion made some time ago--and it became a part of my vision, too--that there be established regional research and treatment centers for HD in major cities throughout the country. The counseling program I'm suggesting would be an integral function of such centers. Skilled professionals provided with the latest information on HD and trained in the most effective counseling techniques would be staff members of these centers. Their primary duties would be to counsel patients and families visiting the center. They would also conduct seminars for specialists in private practice and those specialists attached to public institutions and public agencies in surrounding areas. When I say, "surrounding areas," I'm thinking 200, 300, 400 miles away, in the West possibly. I would hope that this might be true, that they could also train a core of mobile para-professional associates (perhaps volunteers, perhaps on the staff) who could counsel families unable to travel to the centers. I have some confidence that such a program would benefit patients as well as other family members, allowing them to live happier, more useful lives, and to solve more of their own immediate, practical problems. Thank you.

M. WEXLER: Thank you.

GUTHRIE: I want to pick up again on a very important phrase. You said, "realistic assessment." Had you said that ten years ago, we would have said one thing; two years ago, we would have said something else. Tonight we say something else. I think it's very exciting to realize that there are a group of people, whom we call professional counselors, who are learning from us what the realistic assessment really is. It's a question of time, and we must continue in the ways which you've said, including what we're learning from each other.

STARR: If I might add, I think that maybe one of the valuable things about having a counseling program as part of such a system of regional centers is that it would allow some professionals to do firsthand research in just that kind of field, the behavioral field.

M. WEXLER: I think that you had suggested that this could be, in a sense, broader than just Huntington's; that if you had counseling centers, there could be fed into a center of that type problems that involve some 400 other dominant gene diseases, some 2,500 other genetic disorders, a vast array of neurological disorders, movement disorders, and things of this type. Would it appeal to you if there were some kind of model for Huntington's set up as part of a structure into which all the other diseases could be fed so we'd get the highest order professional?

STARR: I'd buy that 100 percent.

N. WEXLER: I think you should know that we have two work groups of the Commission, one in counseling and one to plan these kinds of regional centers that you've been talking about and other people have recommended. We will make sure that these small study groups get the details of your recommendations.

STARR: Thank you.

M. WEXLER: Could I call on Iva Kline, please.

TESTIMONY OF
IVA KLINE

KLINE: I wasn't prepared to speak, so it's not the way everybody else spoke. I hope you'll understand. My life of Huntington's disease first started when I married Albert [Noddleman] in 1948. All I knew of that was that his mother had had some illness. The family told me about what it was, but I kept forgetting the name. They said it was something. As fast as they told me, I forgot it. I asked the doctor before we got married, "Is there any reason why I shouldn't?" He said, "No," so we did. We had three daughters. After my third daughter was born, I noticed that he started losing weight, inflections in his speech, little things. He worked in one place for ten years, and finally they let him go because he was working with bottles of liquor and was dropping too many. After working ten years they had to let him go with no notice of any kind. Just that one day, all of a sudden there he was without a job. I had three small children to take care of and no money coming in. That was one traumatic experience, but as the years went by there were others.

We lived in Chicago. It got very difficult for him to walk, especially on the ice, so we decided to come out to California in 1963. Around 1964 his condition became worse. As I went to work part time I had to leave him alone with my daughters, who were just young teenagers. We didn't have any help at home, and I didn't think I could go into the expense of hiring anybody for him. What I had to do was put him in a convalescent home. I went around and around, it seemed like, in circles trying to find a place for him, which was very difficult. I found one convalescent home which was expensive, but the government paid something at that time. He stayed there for about a year and a half, and it became increasingly more difficult for me to finance. The convalescent home was \$250 a month then, which I've heard has gone up since. It was in the neighborhood, which was convenient. I didn't have to go too far to visit him, which I did about two or three times a week.

Finally, I asked some friends, "What can I do?" I took him several times to Veterans hospitals in the neighborhood and tried to get him into UCLA and Sepulveda. I brought him there and they said, "I'm sorry, we don't have room for him," so we went back home. Finally, after he was still in this convalescent home, this

one friend (who was in the Veterans of Wars) suggested that the only way I could get him into a Veterans hospital would be to literally leave him there and just forget about him, abandon him, which I did one day. I just took him out of the convalescent home and took him to this one, [Selmar], that's no longer there. After a few days they contacted me and I heard that he was being taken care of. That was in June of 1966, and by September 13th he was gone; he passed away. His condition deteriorated very fast. I don't know if it was for the lack of care, or the wrong care, or whatever it was, but that was that.

Now my problems are that I have a young daughter who is just 22 years old. Last June--I think it was June 5th--her behavior the month before was very erratic. She just ran out of the house and ran away. That was the day of an HD meeting, in fact, that my husband was supposed to go to. I had to go work and he was out looking for her, so he couldn't go to the meeting. Anyway, she was picked up by the police and in a very bad state of mind mentally. I didn't hear one way or the other where she was or how she was until about four days later. She was picked up and brought to Camarillo Hospital. She tried to escape a couple of times, and now she's in Patton State Hospital. This girl was the quietest, most soft-spoken--well, I can't even tell you. All she wanted to do was give to charity. People would call me up on the phone to contribute to different organizations and I would say, "Give it to Paula. She'll do it," and she did. She always went around the buildings and did all these things that I couldn't do. She was very giving and very gentle and only wanted to help people.

Now that she's there I feel that this isn't the right place for her. She was showing some signs of HD the last time we visited her. Even though she's very young to have that, I wanted her to be diagnosed to see if that's really what it is because her movements have become sort of violent. The last time we were there she was not so bad; but the time before, they had her in restraints because they said she was lashing out at people. When I saw her like that, I was very upset.

I just hope that there could be some place where she and other HD patients could be put, especially closer to home. Right now we have a very old car that's just barely getting us where we have to go. Every time I have to make that trip out to San Bernadino--and

we live in the San Fernando Valley--it's an hour and a half ride, and we make it there and back with a lot of complications. It's just like an obstacle course to get us there and back. I'd love to go and visit her at least once a week, and I'm lucky if I get there once a month.

...San Fernando Valley, hopefully, where we could have her or other people that have the same thing so that they could be counseled and taken care of in a nice atmosphere, I'd be very appreciative. I thank you very much.

M. WEXLER: Thank you.

GUTHRIE: I just want to check something that you said. How old did you say she is?

KLINE: She was 22 last June. She'll be 23.

GUTHRIE: Can you describe how much involuntary movement she really has now?

KLINE: The last time I saw her she was moving quite a bit unvoluntarily.

GUTHRIE: She did have involuntary movement?

KLINE: Yes.

GUTHRIE: The reason I'm questioning you is because there are many patients in the mental hospitals who do not have Huntington's disease, but they are people who cannot cope with certain realities which they have to face. The way you've described it to me, and your daughter being as young as she is, I am not so sure that it is Huntington's disease. It may be a psychological reaction to the knowledge that she might have it. I'm not saying it as a doctor. I'm just saying it's a question.

KLINE: I had seen different signs in her speech, although she had gone to school up until she ran away. She was taking a course in psychology, to be exact, at Valley College, which was very surprising because in high school she just barely got by. I thought, "Thank God she got out of high school." About a year later I had

taken her to a doctor--I don't know if I should mention the name--and he didn't diagnose her as having it, which I thought she did. He said, "Why don't you take her to a psychiatrist?" I mentioned that to her on the way home that day, and she said, "Oh, no. I'll go to school, and I'll do this." But she has never worked, and she didn't do the normal things a girl of that age would do, like go to work, or go to school, or anything. She just was sort of doing nothing.

GUTHRIE: I just want to comment--and I think it's important to know--that there are people who assume the mannerisms of Huntington's, and then two or three years later we find out that the patient does not have Huntington's. I think I mentioned to you earlier--and I want it on the record--that we know of a particular case in which a man supposedly had Huntington's for nine years and has just been told that he doesn't have it. He had taken on the mannerisms because of the fears and the anxieties. Because your daughter is so young, in comparison with other patients, is why I questioned it. Who knows? Perhaps in time we'll get better diagnoses.

KLINE: What I'm hoping right now is for April Morgan to go out and see her and examine her.

GUTHRIE: That's right.

KLINE: I only said that this is my opinion. I said she hasn't been diagnosed as yet.

M. WEXLER: Thank you very much. Molly Squirrel.

TESTIMONY OF
MOLLY SQUIRREL
BURBANK, CALIFORNIA

SQUIRREL: When people get sick, the first thing that seems to be neglected is their teeth. Since Huntington's patients usually die from secondary causes, and one of the commonest things is choking to death, I think it should be pointed out to the families and the Commission that you should take care of your teeth, first of all, so you don't choke to death (unless you want the person to go extra fast and let it slide). We know that most patients wind up in mental hospitals; and there, all they do is plug in and fill in the holes, or they pull out the tooth when it bothers them, and then it makes it even harder to chew.

My father-in-law died in Camarillo. He choked to death. He was left unattended while eating. So, I think you ought to be aware that you ought to get the best dental care that you can. Even though I'm only a dental assistant, I discussed it with my employer and his associate. We have one patient who has a hereditary neuromuscular disorder, Niemann-Pick disease, and we're trying to be extra conscientious with her and her teeth, because if you have a removable partial and you cough or anything else, and you can't control it, you'll swallow the thing. Also, if you take out the teeth, as I said, you can't chew anyway.

Okay, we know a lot of people say it's expensive and the patient won't hold still. What we recommend to patients who have muscular problems is that there's a general dentist in our town who likes to use--it's cheaper than going in the hospital for all your dentistry--to find somebody who uses nitrous oxide. It seems to relax the patient just enough so you can hold him still and get the work done. It's cheaper. It saves you a few hundred bucks. That's dental care. Okay.

It's not as glorious as John's plan, but I did want to recommend, too, that I think psychiatric counseling programs would be terrific. I know they have them for married couples in Ventura County as low as five bucks a month, and you can go and get this group counseling. It's on a sliding-fee scale. I think if they had the same thing for all these families that I know up in that area with these hereditary diseases and all the attendant psychological traumas, and all that, that that would really be terrific. I think that the state, if they're going to support it for that, can jolly well help support it for this, too.

Jan thought this was interesting, but I just thought, "Oh, well. That's just my weird husband." You need psychological counseling even if you don't have a disease, or maybe you may have the disease secretly but you don't know you do. I think you need counseling whether you get it or not. We were married for three years before I finally convinced him that just maybe things would stay stable enough to where we could buy some furniture. I mean, it was three years, and I would go and window-shop. Finally, we got to buy a couch. Now we've been married nine years, and we just moved into our first house. His common statement was, "Well, we might move, we might move. What if we have to pack up and go suddenly?"

You think, "Man, man. What do you mean, 'pack up and go suddenly'?" Really, people pack up and move across the country without a dollar's worth of furniture. He was always waiting, but he would never say what he was really waiting for. He was lining everything out so he wouldn't have anything to leave in a will, see. It would make things really neat [laughter]. At least now I've got it to where I'll get something [laughter].

Before he was married to me he was engaged three times. Once he really thought he was going to get married to a nurse. However, she had an HD patient in her ward. After watching him a certain amount of time she thought, "Hmm, I guess I don't want to marry him after all," and that really broke him up.

But it shows how it does break up your life whether you get it or not. You have "shrink" expenses whether you get it or not. You wind up going to the "shrink" and saying, "Why won't he let me buy the couch?" [Laughter.] Really. Also, his brother is 42, and he's single. He's been to a "shrink," and he'll never marry. (He once carried on a mild flirtation with a doughnut girl, but his mother found out so that was the end of that!) [Laughter.]

One example of how people seem to be distressed by the disease is that one friend of ours, who thought she was helping, gave us the L.A. Times article when Dr. Wexler first founded this chapter of CCHD around '69 or so. They were invited for the weekend. She brought the article down to the house. (We were renting at the time.) [Laughter.] She put it on the table and said, "Hey, look. I've got this for you. Now there's somebody doing something about the disease." Reaction: "Ahhh," and he ran out into the middle of the street. I couldn't get him to come back into the house while the article was on the table. That is his reaction. Of course, he's come a long way since then.

That is, unfortunately, the reaction of most of the relatives. We have to be aware that these people need psychiatric counseling a lot of times just to have some companionship from somebody who may have the problem, because the relatives won't associate with them. We get a postcard once a year from the cousin. Because he's a teacher and he gets home early, I'm sure that when I write back my nice loving, long letter in reply, he must get home and throw it out or something before his wife gets there (because she's

the one who always sends the Christmas card) to make sure there's no contact established. Plus, he was married for 12 years. I met his wife and she said, "Oh, do you have any kids?" and all that. I thought, "Well, this is my husband's cousin's wife, and I can talk to her." I said, "Well, no, because of Huntington's disease, you know. We probably won't." "Huntington's disease?" (She had three children by this time, by the way.) I said, "Yeah. You know, that runs in their family." "What? What's that?" That's how she found out. That's part of the reason the cousin won't have anything to do with us, I suppose. This just shows some of the trauma. She found out then why her mother-in-law was hidden away in the hills. She couldn't figure out what was the matter with the lady and why they never visited her.

I think they need those counseling groups. I think they need good dental care. If the families have them in mental hospitals and they do want them to choke to death early, don't do anything. If you don't, take them out, go to a dentist and have the best kind of bridgework done or something fixed that won't fall out when you're eating. It'll help you chew. You can feel free to ask me any questions [laughter].

M. WEXLER: You really live and learn. Next time she comes in and says, "Why won't he let me buy the couch?" I'm going to say, "You've got HD in your family" [laughter].

GUTHRIE: I'm very interested in the fact that you are working as a dental technician. We have had a number of people testify and speak of the difficulties of maintaining the well-being of the person's mouth on the whole because of the involuntary movement. It's kind of late, but I would like to suggest again, if we may, Nancy, that this would be a person's brain I'd like to pick further in terms of suggestions for care, because I do believe it is vital and it is something that has been a problem in the care of HD patients. I'd like to make a point of that for management. Can we do that?

N. WEXLER: Go ahead.

SQUIRREL: One of the things we recommend is an electric toothbrush, of course. They can use it more than trying to use a toothbrush.

GUTHRIE: It's more than that. I'm thinking of more than just the patient's care. I'm thinking, for instance, of the patients who have a good deal of involuntary movement, and who might be in a nursing home or hospital where it's completely ignored. I'd like to make some practical suggestions for that kind of care.

M. WEXLER: Thank you very much. Time is running short, and we have one more group. I'd like to get that group up here rather quickly. I'm not quite sure that all the names I'm going to mention are still here. Lisa Eletto, John Hill. Is John Hill here? The next witness doesn't know she's going to be a witness, but she is going to be a witness, Dr. Maryline Barnard. Is Jack Cunningham here?

I'm going to ask Dr. Barnard to begin, and I'm going to do the introductions for her. Dr. Maryline Barnard. Most of you know her, of course. I know that. She and I have been collaborators, researchers for the past 30 years. Our field of research happened to be schizophrenia. At the very beginning of the formation of the California Chapter of CCHD on through the formation of the Hereditary Disease Foundation, Maryline has been a trustee, an officer, a moving force, and the person who does a good deal of our psychological counseling and genetic counseling, organizational work, science board work, et cetera, et cetera. She is also one of those bridges, one of those essential and vital bridges, without which I think no progress could be made whatsoever. She's also connected with the HD Commission since she is part of a work group in the Commission in connection with counseling. Maryline?

TESTIMONY OF
MARYLINE BARNARD, PH.D.
PSYCHOLOGIST

BARNARD: I think you've stolen practically everything I was going to say.

M. WEXLER: You're going to say something about our schizophrenia research?

BARNARD: I think I'd better. I am a psychologist in private practice. Since I have been connected these many years with Huntington's disease I have volunteered to do some counseling for the groups and let people come in. They have been referred by doctors,

especially right after diagnosis. I have seen families for the things that all of you have been talking about, in the way of all the problems that have come up. There just hasn't been any place to send anyone. When Nancy said to me, "Can't you get some psychologists to come and testify?" I couldn't think of one single psychologist in this city that I know of who knows about Huntington's disease, or to whom I could refer a patient. In fact, not long ago, there was an at-risk patient who wanted a referral to a doctor for some therapy. I asked a colleague of mine if he would hunt around. The same answer came back, that there wasn't anyone educated in Huntington's disease to take that aspect and do that kind of counseling with a patient who really wanted some help.

The appointment of April Morgan to the tissue bank--that's why I say Dr. Wexler has already said all of this--she is vitally important. I wanted to stress not just her link with research and with the patients. If we had April cloned a dozen times, we could have an April who met with the at-risk people in a group and just did group therapy with them. In fact, I said this to her one time, and she said, "If you can get me a grant so that we could do some therapy with the patients and with the families at night, I would be glad to do it." I think that that would be a great step for the Commission, to use these at-risk people. You can see the enthusiasm in this group that has been here today. They have talked about these great centers and the great professional people, but I have found in my experience that it's an at-risk person who can help another person who is at risk if they can just sit down and talk together. If we had a grant where professionals could train the paraprofessional at-risk person to then go out and get groups together, you could accomplish a great deal in a very quick time.

You have to have a person who can give full time to it. I don't work out because I have to work for a living, and what I can do is just a spit in the ocean, and it's not enough. If there was somebody who did have time to go out and really organize the at-risk people and the families and the Mrs. Geers who know about the problems of the wives and the husbands who are at home taking care of the family, they can contribute a great deal.

Being a therapist I know that no therapy ever takes place unless the person is involved. I think that you people are the ones, if you are given some kind of professional help, who could really

get involved and help yourselves. That was the recommendation I wrote to Dr. Falek, who is head of the work study group, that he consider a budget for training selected at-risk members of the HD population to organize or lead groups, or even man a hot line (just a telephone thing), or organize day care centers. For example, for a city the size of Los Angeles we need key people that are spotted around in various parts of the area who can be reached in emergencies, who can educate the community, and who can research community facilities so that they could tell people where to go for a convalescent home. This could be a full-time job and should be reimbursed in some way, possibly as grants to the voluntary organizations that are equipped to train the personnel and supervise the work. I'd just like to add that in addition to all of the other very fancy centers, and what not, that have been recommended before.

M. WEXLER: Are you suggesting in a more general way, apart from the question of the at-risks and the families involved, that it would be very useful for some group of educators to develop curricula (let's say for clinical psychologists, for nurses, for psychiatrists, for neurologists, for medical schools in general) of various levels of intensity so that all of these populations that deal in a professional way with the problem of HD would have available to them the materials to incorporate something about HD in their more general curricula, and that it should be really geared at these various levels, for nurses, psychologists, neurologists, et cetera?

BARNARD: It would be an excellent place to start. What I wanted to emphasize was that it would be this group [at-risk people] that would be in there and would be trained in some kind of way to go right back out to their own people.

M. WEXLER: You wanted to emphasize the paraprofessional aspect of it?

BARNARD: Yes.

GUTHRIE: I can say that I do agree with that as a possible method for helping people. I think even today we saw how comforting it is when one person meets another patient and they work around the fact that they share a mutual problem. I did attend a meeting where there were four men, all in the middle stages of Huntington's. When the tea and coffee were served at the end of the meeting, those four men found each other. It was a fascinating thing for me to watch and

to see how it seemed to help them. I'm sure that your suggestion is that that person could even do a better job with some professional guidance and training.

BARNARD: Right.

N. WEXLER: There's a big movement in the country now of volunteerism and self-help. In fact, someone is writing a paper for the Commission about self-help in HD. I think everybody assumes that everybody has time and resources to do the self-help. I think what you're suggesting is really critical, that there has to be some kind of funding or seed money.

BARNARD: Right.

N. WEXLER: If you expect people to help themselves, then they have to be able to take time out from a job to do it. Considering your own training and all the work that you've done with HD families, do you think there should be some kind of training for clinical psychologists or counselors or social workers in helping people deal with either chronic neurological or genetic diseases, that there's a certain body of information that you just can't expect every counselor or psychologist or psychiatrist to know by virtue of going through their training program?

BARNARD: Correct. The colleague of mine who was trying to help me find someone said he was going to go right to our L.A. County Psychological Association and bring it up, because he thought that there was ample opportunity there to do a lot more education for our profession in that area.

N. WEXLER: I would just like to say one thing about you. I've been getting special, privileged counseling for all of these years of my life. You're getting treated to my whole family. Maryline is my surrogate mother. If we could clone you, I think that a lot of the psychological problems of all the people in the room would disappear.

BARNARD: Thank you.

M. WEXLER: I don't know whether you people appreciate the kind of impact that this statement can have on a person like myself.

When I think about it--right up here there are three people who have gone through any number of college degrees to get training to do very advanced clinical work in psychology--I'll bet that not one of us has had a course in counseling anybody with a genetic disorder. I've had 4,444,000 courses (educational, clinical, otherwise), and so has Nancy and so has Maryline. Not one of us has had a course on anything to do with hereditary disease, genetic disease, its impact and the psychological status of patients, et cetera, et cetera.

If you think that's sad, I think you probably would find that the same is true in medical schools, with some few exceptions. These medical doctors come out with their advanced degrees, and it's not just that they haven't heard of Huntington's disease; they haven't heard of the fact that people with hereditary disorders, degenerating neurological disorders have some kind of psychological problem, which strikes me as an amazing gap in human knowledge and an amazing carelessness in the development of curricula. Lisa? Lisa Eletto.

TESTIMONY OF
LISA ELETTO
LOS ANGELES, CALIFORNIA

ELETTO: I like to do things chronologically. The story, as I heard it from my grandmother, was that she didn't understand what had happened to her husband, who was normally very gregarious and outgoing with a real good sense of humor. He was just a changed man. She didn't know what the disease was. She didn't know it was a disease. They divorced when she was about 35 or so. Looking back, this must have been really a hard thing for her, coming from Nebraska (where you just don't get divorced), uprooting and coming out to California. I see her now, helping to care for my mother. It's just a lifelong struggle, as I see it.

As a child I blamed my mother for her temper. I felt that she could control herself, and I didn't realize that it was uncontrollable. In junior high school I remember reading about Huntington's disease in one of the textbooks. They devoted one sentence to it, and the sentence was rather alarming. They felt that Huntington's at-risk people should not marry because of the risks, and this was my first exposure to it from a professional angle. Then when I got in high school it suddenly hit me that I was at risk when I was about

16 years old. I'd been so self-righteous about it before, like "Why can't my mom control herself?" and so forth. It just suddenly hit me, and I was just plunged into depression for about six months. I didn't know that other people were at risk, that other people shared the psychological problems. My father also experienced an emotional breakdown because of the stress. That was when it first hit really hard. Adolescence is a hard time to go through that, and I know a lot of people with parents who have HD probably go through these same experiences at about that age.

When I got into college the dating scene arrived. For one thing, the friends I talked to and confided in told me, "Oh, just don't tell the boys anything about it. Even if you go steady with someone, don't tell anyone, because you won't get it." Even at a young age that just struck me as complete nonsense. On the other hand, my policy of honesty was really hard to take, too. One boy friend went with me for a year and a half. When I finally got around to asking him, "Hey, do you think there's any possibility we'll ever get married?" he told me that the reason he had gone steady with me was because he knew I was safe; he knew he would never marry me because of Huntington's disease. It just, wow, sent me for an incredible loop. Once again, six months of depression.

The next boy friend just avoided the subject. I was open about the disease, but too scared to probe his ideas about it. It was only through my friends that I later learned he was scared to death of it. The third boy friend was a medical student, and we talked openly about the disease. He had had some HD patients in the hospital and had seen what they were going through. That kind of scared him. Also, the senior doctors talked to him about it and advised him not to marry me, that it was too heavy a burden. I feel pretty fortunate that I met a man who loves me and accepts me. It's just like the biggest difference in my life.

These were the antiwar years. I remember thinking during college that being at risk for Huntington's was like being 1-A for the draft. It just struck me that way. It's just always hanging over your head. Every time I got depressed about one thing, Huntington's would enter into the picture. Psychologically, I know I blamed a lot of my problems on Huntington's disease. For some reason I guess I had an idea that I had to be strong, that therapy was for weaklings, or that it was not acceptable. It was quite a bit later that I did go into therapy.

I was open with my friends I met on my job. I'm a teacher. However, I've always been a little reluctant to confide in my boss that I am at risk. I just feel if I continue to teach there for a long time, maybe they'll see things that they regard as symptoms. I just don't want people looking at me that way. It's still a paranoia I have.

After a couple of years of teaching I entered the counseling program, the M.A. program, at Northridge. It was only in that program, where I was exposed to a lot of group counseling and involved in it myself, that I suddenly realized I really did belong in therapy. It opened me up and made me understand that I had a lot of problems that couldn't be solved by going to class once a week and talking with some people. The therapy was incredibly necessary. I went through two years of therapy, and I feel 100 percent better about myself. I feel I've accepted the disease to a much larger degree. I can't say totally. I don't know if anyone can say that.

I have grown much closer to my mother emotionally, identifying with her and thinking back to the memories I had of her as a child, looking at pictures, talking with people, that kind of thing. It was very important for me to have that identification with her. I learned to give up my resentment against my parents for the way they handled their problems with the disease.

At the point I am right now my husband and I are both hoping that some day we can have children. For a long time I felt I didn't want to have natural children if there is a 50-percent chance of them having HD. I'm looking at the research and reading the little pamphlets and hoping that eventually they will come up with a test that can determine whether I have it at least. If I found out that I did have it for sure, I wouldn't have my own kids; of course, if I found out I didn't have it, I would. That's the issue right now in my life.

In regard to my family, I've noticed a lot of people talk about my mother and not at her, which disturbs me. I know I did it for a long time myself and sometimes catch myself doing it. The person with HD is sort of regarded as not there, and that's disturbing. My parents have now hired a part-time nurse. They would like to hire a full-time nurse, but it's kind of difficult. We were lucky enough to have Travelers insurance, because my dad

had a group plan through his work. If his work had not had a group plan, it would have been almost impossible to get insurance and medical care.

I have a sister who is 17. It's going to be harder for the family now that she's gone to college and is no longer there to care for my mother during the day. She personally has felt that she may commit suicide if she gets the disease. When I heard that, I started evaluating it myself, and I just don't know where I stand. I hope that life will still have enough meaning for me to go on with it. She remembers as a child being teased by a friend who thought mom had drunk too much, and that was a pretty traumatic thing for her.

The one thing that impresses me most about the disease--it's overwhelming for me--is that it's so debilitating, not only physically, but socially. People just don't understand what it is, and you become sort of an outcast from society. People stare at you, and they don't understand what's wrong. Even when they do know what disease it is, it's sort of like you have the plague, or something. It's very hard to handle, and I think that's one reason people need more counseling, to learn how to deal with it and not avoid it, or not become patronizing toward the person who has the disease. Last year my mom and I went to a restaurant. The hostess said, "We can't serve you. She's drunk." I went into my big, long spiel about, "Well, she has a disease called Huntington's disease," and so forth. She said, "Are you sure?" At that point I said, "You can take your restaurant..." and left [laughter]. It really, really upset me more than I wanted to let on to her, and I was really proud of my mom for the way she handled it.

There was one more point I wanted to make, and I guess I made it at the beginning. It seems like people get hit twice with Huntington's disease. It's something that a person should only have to endure in one aspect during their life, but most people are hit twice: first, as a child growing up with a parent who has Huntington's disease, having the problems of dealing with that parent; then being at risk and all of the manifestations of that, all the psychological trips you go through; then you either get the disease and have the disease to deal with, or you don't get the disease but usually have a parent or a sibling to care for who has the disease. So you're really hit twice, and it's a double burden.

The needs that I see are more medical insurance (possibly subsidized by the government) and nursing home insurance; more research; hopefully a test to determine who has the disease; treatment, so that if you do have the disease it's not so bad; and, as so many people mentioned previously, counseling. Every single one of us needs it at all stages of the disease, at all stages of being at risk. I remember the first HD meeting I went to that was just for people at risk. I was in college. I just sat there, and I couldn't believe that all these other people were at risk. I wanted to touch them, and I wanted to say, "I can't believe I'm not alone." It was just overpowering. I think it would be great if we had peer groups for people at risk and people going through different phases of the disease. Thank you very much.

M. WEXLER: Thank you very much, Lisa. Mr. Jack Cunningham, please.

TESTIMONY OF
JACK CUNNINGHAM

CUNNINGHAM: My name is Jack Cunningham. This is the first meeting I've ever attended, and it will not be my last. I've learned a lot already. My wife is 46 years of age. In 1973 we learned that she had Huntington's chorea. I think I'm good, but a late starter. My children are from 15 down to 8 years of age. I'm concerned with my own health so that I can take care of the gang as the years go on.

Because of all the doctors and psychologists and scientists that came here and are interested, I'd like to tell you of the good experience I had. We went to our internist, who said my wife was physically perfect, but nervous. He recommended a psychiatrist, and we went to see a psychiatrist for maybe six or seven visits. He mentioned that her movements were almost Parkinsonian in nature. He said, "Before I do any more, I think you should see a neurologist before I go into shock or anything more serious." We went to see a young neurologist whose father had been a plastic surgeon. He said, "I think I'm being very brash--he just looked at my wife this one visit--"but I think your wife has Huntington's disease." I went to the local bookstore and looked it up, and that shook me up a little bit. I told the psychiatrist, and he said, "Well, he was indeed brash, because there's only a 15-percent chance of that." Then we

had a neuroencephalogram made. I was going to go to U.C. instead of St. Joseph's, because "We're going to have the best." The psychiatrist said, "Now you're overreacting." She had the test. It turned out that in this doctor's opinion, with the frontal brain horns and pictures and things, that she did have it. He sent us to a doctor at the University of California at Irvine, or Orange County Medical Center, a neurologist who has turned out to be a dear friend of ours and who confirmed it. He also recommended to the other neurologist that "I think I can take better care of your wife down the road in this particular area." (This man has been written up in Newsweek and a few magazines.) In '73 we went to New York City on vacation and went to see a doctor at New York University who confirmed the diagnosis. It turned out that he went to high school with the U.C.I. man. I really feel empathetic to the people that have gone 20 years. At least I was very fortunate in that I think the psychiatrist was--I owe him a great debt of gratitude to do what he did.

When it first started out it looked like a nervous breakdown. She was afraid she was going to make ethnic slurs. I worked for a German company, and she thought she'd call people "Nazis," or "Italians," or what have you. She was a small lady, about 105 pounds, and she blew up to 120 because she hated housework, and what have you. We coped as best we could, and I had an empathetic boss. In 1974 she put her hands around my five-year-old's neck at the breakfast table. I'd like to mention for the record that, right or wrong, we have not advised the children of anything, except that their mother has a neurological illness at this point in time. Her relatives find it difficult to even recognize that. They thought it was 15 years with me, but they're finally learning that it's more than that [laughter]. I love her, and I'm going to hang in there with her, so that isn't it.

She went to a regular hospital for 23 days and had all kinds of tests. They did bring in all the interns and psychiatrists at Orange County Medical Center so they could learn: "Young fellow, you recognize this when you see it in your practice in future years." Then the doctor said, "Jack, I don't think she'd be any more unhappy in a hospital." I said, "What do you mean? What happens after the \$50,000 major medical runs out?" He said, "Well, it's a state institution." I said, "How are they?" He said, "Well, they're not perfect. I suggest you walk through one." I think he was thinking

of the five of us versus one. I said, "No, I can't do that." I had no relatives out here and the kids (the oldest was 13 and the youngest was five at the time), so I sent her to Brea Neuropsychiatric Hospital for 30 days, and the kids and I hacked it on our own. There she met a psychiatrist in group therapy who had been a Navy surgeon at one time, a young chap. She got into group therapy, and she found so many people--like, there were Quakers who were out of their heads because they had killed people in Viet Nam, pregnant girls whose husbands had left them when they were 17. Anyway, he said to her, "Look, honey, you're not out of your mind. You have a physical illness that destroys the brain cells and affects the limbs." (The neurologist told her she would not live a normal lifespan.) He got her out of it.

We brought her home, and we had this nurse who was about 26 or so, and a fun kid. She came over in her starched uniform the first time. The next day she came over in shorts, with our permission. She used to do more than nursing. She'd make cakes, and she'd take my wife to Laguna Beach to go through the shops and to the bank. She got her back into the mainstream of things. We continued the psychiatry and she progressed. Then she stopped the psychiatrist and went back to the doctor. He said, "How are you feeling?" and she said, "Good." She was on Artane and different things. He said, "That's pretty heavy. Let's cut it in half," so we cut it in half for six months. We went back, and he said, "How are you feeling?" She said, "Good." He said, "Let's cut out the medication entirely," so she's been without any medication for a year. She had been on Maldol. She's doing her three or four loads of wash a day. She yells at the kids more than she used to and has many of the things. Boy, am I windy.

I don't know what religion he was, but part of this psychiatrist's technique was the "supreme being." My wife practices her religion, as does the family. She has strong religious convictions, and she gave up black coffee, martinis and cigarettes cold turkey as an offering to God to make her better. (I haven't given up any of them yet) [laughter]. In fact, I'm using them--well, we won't go into that one [laughter].

I think I have some nice, attractive young children. My 16-year-old (who will be 16 May 31st) did say in a fit of anxiety one day that if mother's illness was hereditary, she would kill herself.

Getting back to the denture problem, you have to remember how fast an illness can come on. She had the last child in '69. In '73 we moved the five of us from New York City to San Francisco. She had periodontal problems for years, and we spent a lot of money on them. Finally, she lost, first, her uppers, which she can handle beautifully, and then the lowers. They don't fit good, and we've gone to several dentists. They fit as good as her gums will allow. The dentist told us that the lowers require muscle control. So, if you have to lose any, I guess it's better to lose the uppers first.

...her dentures out fast. When she goes to the altar at the church we go to, I take them out, the lowers, when she goes up to receive the Host. That frightens us a little bit. That dental thing is an important aspect, in my opinion.

My concluding comment is that I refinanced the house and a few things. We thank the Lord we're in Orange County where things are appreciating more than in most places [laughter]. My concern is my ability in the next number of years. I'd like to do the best I can for her in a home facility. There are no relatives or big affluence on either side of the family. When I think of the cost of a mental institution, if the government (through Social Security) could have some kind of--like, if she had worked five of the last ten years, she would get Social Security. I wrote Mrs. Guthrie a letter about this. I appreciate your answering it. If she had worked--but she quit in 1957--she and the children would be getting Social Security, which would be maybe \$600, \$700, \$800 a month, which would allow us to do more things.

In my business, which is the automobile business with an automobile factory, when we put a dealer in a specific location, the three requirements are retail automobile experience, adequate capital, and basic integrity, meaning he's not a Mafia member, or something [laughter]. I think we have the experience, but we're undercapitalized to take care of our loved ones in some respects. Doctor, I'll conclude. I fall into that category of had been happily stuck in the mire of middle management, doing fine; neither rich nor at the poverty level. That's when you're on your own, sir. You summed it up very good. Thank you for your patience, everybody.

M. WEXLER: Thank you. I have a new definition for the word "courage." It's someone who faces the plagues of Job with a sense of humor [applause]. Mr. John Hill.

TESTIMONY OF
JOHN D. HILL
NORWALK, CALIFORNIA

HILL: Thank you, Dr. Wexler. I'm glad to take this opportunity to testify before the Commission. I'm grateful to you, Dr. Milton Wexler, and you, Nancy, and Dr. Barnard for all the help you've been in the past.

I've been associated with Huntington's disease for approximately 30 years. My wife has Huntington's disease and has so been diagnosed. Her mother had Huntington's disease, and she tried to care for her to the best of her ability for a long time, but she was unable to.

We have two children, a boy (22) and a daughter (12 years old). We also have her brother, who has lived with us since he was six. His father passed away of a heart attack, so we took care of him and raised him. These children are at risk. They know it. They've been explained the counselings of genetics and also about the possibility of being at risk.

We went to several doctors (psychiatrists) for counseling and marriage counseling over the years. Most of it was fruitless. As things progressed, my wife was diagnosed as having HD. This was quite a blow, even though it was suspected. We are very fond of her and we love her very much. We hope that something can come from this meeting to help relieve it.

I guess we've all experienced in one way or another the various aspects of how people who have HD relate. One very important thing we find is that they are sometimes unable to relate to reality, and those of us who are also associated with it find it difficult to relate to reality, too. This comes to the point of this counseling, psychologists. I think it's a wonderful idea for all of us if this could be brought about to offer help somehow, to stabilize our lives, to live more fruitful and happy lives. As to what Lisa said tonight, I wish she would write this down, so I could present her story to my 12-year-old daughter. It's very important. The things that she said would help us all.

We found most of the people who have HD to be very charming, very lovable, very kind, and very generous at one time; otherwise, we wouldn't have been attracted to them. To have them in a home by themselves, off to one side is not our desire; but to have them be with everyone, wherever they can, for as long as they can. I wouldn't ask for a special place for them, because they have the feelings we have, and they would love to be around us as much as we would love to be around them.

In conclusion, I would just like to reemphasize the need for the counseling. Each and every one of us could possibly be of help to one another in the realities of life with a little skill and a little more knowledge. We'd appreciate it. Thank you very much.

M. WEXLER: Thank you, Mr. Hill.

N. WEXLER: You actually can have what Lisa said, because we transcribe what's been going on tonight and in nine other places where we're having hearings. After these proceedings are published, all of you can have everything that's been said all around the country. You just have to let our Commission office know, and we can get it to you. You can make sure that the right people read it.

HILL: There's one thing more I would like to say. I didn't have any idea that my wife would be here tonight, but she is here, sitting over here.

M. WEXLER: It's awfully late, and we've overstayed our leave. I have a feeling that there are a great many people out there who would still like to say things. I think perhaps, however, we had better call it an evening and call it a testimony.

I did want to take a minute, rather selfishly, to first express my appreciation for everybody coming here, for so many of you being so patient for so long, listening so carefully, for the contributions you have made in terms of your dedication, your interest, and your involvement, and to tell you a story which just came to me this moment. I don't even know whether I can remember all the details. I don't know the ending of the story, but I have a hunch it's in some way relevant:

I'll tell you the story about a little town in Belgium that was really built up around the thirteenth century--oh, I know why this story comes to me. It's very embarrassing. There was an Irish king who had a daughter, and she was a princess. There was wild jealousy that existed because he didn't want his daughter to belong to anybody else, and she wanted to get married. There was a priest, and he listened to her story and thought that the father was somewhat mad, which he was. At the pleading of the daughter, who wanted to be rescued from this tyranny, this awful father, he secreted the daughter out of Ireland and brought her to Belgium, to a little town called Gheel. The wild and mad Irish king traced them to Gheel, and there ensued a tremendous fight and the king killed the priest. He was quite mad. The townspeople, who recognized his madness, instead of revenging themselves, made a vow--it was a strange vow--that they would take in all madmen and welcome them as members of the family. From that time on the people of Gheel have never, never turned aside any schizophrenic who came to the town to seek refuge, safety and self-protection.

Not only that, they didn't hospitalize them. They allowed them to live in the families. It's a tradition in Gheel that all schizophrenic patients are permitted to live within the families; they work if they can, or they don't work; they are loved members of the family. What's really remarkable about it is that because the doors of the town are open to these desperately ill people, these desperately ill people are quite remarkable in their capabilities, in their integration in the family life, in their integration in the work patterns.

If you go out to Patton State Hospital, where a good many of the MD patients land, or Norwalk, or Camarillo, and see the deterioration that takes place when this kind of isolation and treatment are part of their regimen, and then see what happens in terms of the MD patients who are in this sense still members of a family, still visited, still cared for, still attended, et cetera, and you see the differentiation, you know, you really know, that if we are not frightened by the disease, if we are not rejecting of the disease, and if we are not rejecting of the people who are sufferers, but are wide open and accepting and loving, there is a tremendous therapeutic effect on this.

I mentioned it because I think there are some wonderful people over there who talked very tenderly and very warmly about the sufferers in their families, and it's been happening all day long.

Here at the end came to me this rather ancient fable about how it was that in the thirteenth century this wonderful town in Belgium was established.

Apart from any Congressional program, and apart from any funding, centers, research and everything else, I hope that all of you keep your hearts open, and your minds open, and your feelings open to the people who are sufferers, the people who are at risk, and the people who are endangered; that we are, as has been mentioned many times, one large family, and we've got to stick together. Thank you. Thank you for coming [applause].

GUTHRIE: This Commission came about because people like you wrote letters and helped us get the ear of the Congress. This is only half of the battle. The report to the Congress is due in August. Nothing will come of our report and our recommendations unless you become active participants, in a sense selling what we decide are good recommendations to your representatives in Congress. Let's not lose our heads. We're going to finish our job. I think that our staff is beautiful. They've done a wonderful job. The members of the Commission, as you've seen, are concerned and very dedicated people. But we need your help when the time comes to literally sell what we say to the Congress.

I would like to end with a very interesting statement. I have been going around the country, saying, "Life is a fatal illness. Life is a fatal disease." Just two nights ago a young doctor said to me, "And being born is the first symptom."

M. WEXLER: Of course, we're all at risk, you understand.

Los Angeles, California

April 15, 1977

REGINA E. ACHZIGER
PACIFICA, CALIFORNIA

MARCH 5, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

MARY HUGHES ALLEN, M.D.
MENLO PARK, CALIFORNIA

MARCH 29, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, and their families and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

DIANE ANDERSON
MODESTO, CALIFORNIA

FEBRUARY 14, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

JEAN ANDERSON
MODESTO, CALIFORNIA

FEBRUARY 14, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future families, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

RAY ANDERSON
MODESTO, CALIFORNIA

FEBRUARY 14, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

MRS. SCOTT ANDERSON
BERKELEY, CALIFORNIA

FEBRUARY 25, 1977

May I please urge the Commission to make every effort to combat the effects of Huntington's disease (including compensation to families) and to provide research that can lead to a cure.

Having just watched the slow disintegration of a person with Huntington's disease, and the enormous pressure it placed on her family, I feel it is important to try to prevent this tragic illness which keeps affecting families through children of victims.

Los Angeles, California

April 15, 1977

BONNIE A. AUSTIN
SAN DIEGO, CALIFORNIA

MARCH 21, 1977

This month a brilliant woman and friend died of Huntington's disease after years of suffering. Her children now approach the age when the disease will show, or hopefully prove to be uninherited. Without an available test to show its presence or absence, they have lived with the knowledge that it might display itself during their adulthood--now, at any time. And if it strikes, they wait for a cure.

Please make every effort to combat and cure Huntington's disease.

Los Angeles, California

April 15, 1977

DAVID N. AUSTIN
SAN DIEGO, CALIFORNIA

MARCH 21, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, please make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

MILDRED B. BADE
GRAEAGLE, CALIFORNIA

MARCH 20, 1977

I have personal knowledge of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations.

My husband, Allan, died about five years ago after a gradual deterioration that lasted over 10 years. No individual should be subjected to this slow death if it can be avoided through research.

We have two children--one 33 and one 29 and, of course, for their sakes am extremely concerned and hopeful that the next three or four years of research will find the answer to a cure or at least control, so they may always be physically able to support themselves and not be a burden to society.

I strongly urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

ANNE BATEMAN
OAKLAND, CALIFORNIA

MARCH 27, 1977

As a volunteer with no other concern than compassion for others, I beg you:

1. To establish centers for research and custodial care for H.D. victims at minimal or no expense to them;
2. To investigate the problems attendant upon insurance companies who refuse to aid people holding their policies, who are afflicted with H.D.;
3. To pass legislation allowing civilians to participate in these V.A. hospitals without sufficient military patients with H.D. to do significant research--as at Palo Alto, California V.A. Hospital.

Los Angeles, California

April 15, 1977

M. A. BEACH
OAKLAND, CALIFORNIA

FEBRUARY 28, 1977

This letter is to inform you of my personal experience relating to H.D. I was not personally afflicted with this disease but took care of a personal lady friend who lost her mother and one brother from this malady and knew that her time was limited.

I did my best to help until it was necessary for her to enter a convalescent hospital. I made two trips daily to visit her and please believe me when I say that to watch someone whose body is being torn to shreds with their mind intact and knowing what is to happen is the worst experience I want to happen. The Lord above gave her rest so her troubles are over.

Remembering her and knowing that there are many others afflicted in the same way, I most certainly ask that immediate Federal funds be made available to seek out the causes for and the stamping out once and for all of Huntington's disease.

Please open the valves which will start these funds flowing to those doctors and scientists who are seeking the answers.

Thanking you in advance.

Los Angeles, California

April 15, 1977

BETTY BELLFLOWER
FRESNO, CALIFORNIA

MARCH 22, 1977

I am engaged to a young man whose grandmother had H.D. and I am very interested in anything that can combat or cure the disease. I feel that any Federal funds that are available for this type research be allocated to include Huntington's disease.

Los Angeles, California

April 15, 1977

DEBBIE BELLFLOWER
FRESNO, CALIFORNIA

MARCH 22, 1977

I have a sister who is engaged to a young man whose family is afflicted with Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to secure a program of federal funding in order to take steps to combat and cure H.D.

Los Angeles, California

April 15, 1977

MIKE BERTONE
FRESNO, CALIFORNIA

MARCH 22, 1977

I am a very close friend of a family who has H.D.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

JEAN BIER
FRESNO, CALIFORNIA

MARCH 22, 1977

My grandmother had Huntington's disease. My mother is at risk and I feel that in order for families to better understand and cope with the emotional, as well as the physical aspects of H.D., it is essential that every possible measure be taken to research the cause and cure.

There is, in my opinion, a desperate need for Federal funding of Huntington's disease research, as with any tragic disease. However, as a member of a family that has come face to face with Huntington's disease, and having learned that the medical profession has not even touched upon the surface of a cause, much less a cure, I feel it is imperative that your Commission make every effort to combat and cure H.D.

Los Angeles, California

April 15, 1977

J. HARMAN BJORKLUND, M.D.
OAKLAND, CALIFORNIA

MARCH 29, 1977

February 12th of this year ended many years of mental and physical torture for my wife, who died of Huntington's chorea on that date. I can only guess what sad, frightening, upsetting thoughts she must have had in her last years since the illness destroyed the control of her muscles of speech, but I know the hell it has been for my children, my friends, and me to watch a loved one go through such an experience.

But why bring the United States Government into this? Fortunately, I was able to keep my wife at home with excellent nursing care up to the last six days of her life. Basically, my philosophy is that one doesn't cry for help from the Government unless the problem is too overwhelming. For most Huntington's disease families, this is the situation! In my position as President of the Northern California Chapter of the Committee to Combat Huntington's Disease, I get to see that for most Huntington's disease families the psychological, emotional, and financial problems are too great for them to handle alone. They need help from the United States Government!

It is my belief the Government should give some direct help for people currently ill with this vicious disease; but perhaps the best help should be generous support of basic brain research, as well as specific research into the cause of Huntington's chorea and the best medications to treat individuals who are attacked by the disease.

Los Angeles, California

April 15, 1977

PAT BLOMQUIST
REDLANDS, CALIFORNIA

MARCH 31, 1977

I have known Ruth Krueger for 20 years. She was a healthy person most of this time. I have also known her mother and watched her mother die with Huntington's disease. Ruth's become a very different person since this disease has started to take over her body. It is quite difficult for her to even visit with friends. I would hope that necessary funds be allocated to help in finding the cause of this terrible disease. Ruth Krueger lives in Torrance, California.

Los Angeles, California

April 15, 1977

MRS. VIOLET BOGAN
OAKLAND, CALIFORNIA

MARCH 5, 1977

After about 15 years of suffering from Huntington's disease, a friend of our family passed away. A beautiful person and the whole family was so good and kind to her; but they were so helpless to do much for her.

So I urge this Commission to try more publicity on this disease. So few people know anything about it.

Los Angeles, California

April 15, 1977

ANITA BOQUIST
SAN JOSE, CALIFORNIA

APRIL 4, 1977

My father-in-law has Huntington's disease familial and he is about 60 years old and in a veterans home for life. My husband and I are in our mid 20's and we have both seen him go downhill very fast, and it's a very frightening experience to have to go through, and a wife of a husband who may some day have the same disease. As far as doctors who are familiar with the disease there are very few, until lately we discovered Dr. Davis at Palo Alto Veterans Hospital who seems to be experimenting in this area. In my father-in-law's case his intelligence seems to be as sharp as ever, which is really sad in this case since he understands what is happening to him and he is very frustrated with himself and has wished to give up many times. The care he needs is a 24-hour nurse for his medications and care, which is why he is now in a veterans home.

My husband and I have just now discovered the Committee to Combat Huntington's Disease, Inc. in Oakland, California, and therefore we will do anything to help this organization or any other in any way we could to help. It's a very frightening feeling to know my husband may some day have the same problems, and this is why we will not have a family and not spread this to more people for our sakes and the children.

I would like to see more research programs set up for this disease and more campaign programs to make the public aware of this. I was really surprised how many people don't know anything about it, like our family didn't because no one was afflicted. I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

BESSIE L. BOWEN, R.N.
EMERYVILLE, CALIFORNIA

MARCH 18, 1977

It is encouraging that national attention is now being focussed on Huntington's disease and its consequences.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

ANDREW HERNDON BRADFORD
SANTA MONICA, CALIFORNIA

MARCH 28, 1977

My father, Don H. Bradford, had Huntington's disease for more than fifteen years before it was properly diagnosed. The saddest part of the whole experience was that we suspected my father, a man of resilience and strong character, of succumbing to a type of old woman's hypochondria. His numerous unexplained symptoms (vision problems, a nervous stomach syndrome, uncontrolled fidgeting, various aches and pains) that developed prior to the period when he became obviously disabled were extremely deleterious to and eventually destroyed his professorial career, his social life, and almost our family.

An effort should be made to educate the public and the medical profession so that Huntington's disease and its symptoms will be more easily recognized. And, obviously, there is a dire need for additional support so that Huntington's disease and other similar tragic afflictions can be eradicated through research.

Los Angeles, California

April 15, 1977

ETHEL W. BRADFORD
HERMOSA BEACH, CALIFORNIA

FEBRUARY 13, 1977

I received your letter of January 27, 1977, and I will try to answer as clearly as possible.

It is difficult to pinpoint all the problems connected with H.D. in regard to the family and the patient. They are so numerous and endless, to evaluate them, even partially, would consume too much time, and might even be confusing, so I will try to tell what seems most important.

My name is Ethel W. Bradford. I live at the above address. I am the mother of two children, Don H. Bradford, professor, and June A. Clark, both of whom developed H.D. The first symptoms appeared when my children were in their early thirties. My son, Don, a sometimes author, college professor, and brilliant man, seemed to be struck harder, at what was the peak of his career. He was ill for about 15 years and passed away one and one-half years ago at the age of 52. My daughter is still with us, but is only a skeleton. Her mind is just about gone. It would appear she would not last much longer (but this is just guessing). She will soon be 59 years old.

I moved into Don's home and stayed with him the last five years, as his wife had to go to work. I helped with his daily needs, which was not hard. He was a beautiful character. His mind remained clear up to the last day of his life here on earth. He was loving, kind, and considerate of others, grateful for every kindness shown him, easy to be around, gracious, never bitter, even when he found out what was wrong. The last years he couldn't read; I read to him and he was always grateful. He was a pleasure to be around. We all miss him terribly. It was a great loss for his family, the community, and his students, who also loved him. He left three children: Drew Bradford of Santa Monica, Judy Rini of Huntington Beach, and Dulcy Rimac of Valencia, and four grandchildren. Even while he was ill, he saw to it that his three children got a good education, and they are all college graduates.

My daughter, June Clark, has reacted differently to the disease. It affected her mind, right from the start. I could

go on and on about the problems concerning her, that her family has had for years. I will just give the highlights of these problems. Her husband died five years ago, and she has two teenaged children, Amy and Alan Clark, 17½ and 19 years old, whose lives have been made terrible. She started developing this disease when they were babies and, of course, we did not know what was wrong. They have never known a normal home and love from parents.

Since my son's death, I have moved in with my daughter and her two children, and I try to help all I can. She is very difficult to care for, makes it difficult for everyone, refuses to eat (she can only take liquids, cannot swallow solids), and would just sit and starve if she was not constantly urged to eat, is fault-finding, cross, does not sleep at night and keeps others from sleeping, so no one wants to be near her. She is a complete stranger. The daughter I knew and loved died mentally 20 years ago. Her two children's lives have been so warped, their outlook on life is not good or wholesome. They have the hopeless feeling that they have a 50% risk of having the same terrible disease when they reach maturity. I am emphasizing how this disease has touched the lives of these two children and almost destroyed them. I will not go into detail.

Now, in regard to the questions your letter asks. Question 1 - Being "at risk" for H.D. is the fact that an "at risk" person has the risk hanging over him of developing the disease at maturity, and should not plan a marriage, or children, or to lead a normal life, so the world says. There was an article in the L.A. Times a couple of years ago, about a young man, when faced with this, jumped off a Holiday Inn building up on Sunset Boulevard.

Question 2 - The problems and needs prior to diagnosis, is to me, the most important, amazing and terrible ignorance of the medical profession. Prior to the correct diagnosis of my son, Don's, illness at UCLA, I cannot tell you, words fail me, what he and his family went through for about ten years with M.D.'s, psychologists, psychiatrists, neurologists, etc. (the whole gamut of so-called "brains" in the professions). They diagnosed him all the way from a psychosomatic, neurotic, alcoholic, etc. For instance, my son went to one psychiatrist for help. When my son sat down across from his desk the doctor leaned back, rolled a cigar and asked, "Whom do you hate?" Don said, "I do not hate anyone. I love my family and my job." The doctor said, "You must hate someone, or you would not be like this." He charged Don \$40 for this visit.

Another doctor (who represented the insurance company, when Don was trying to get disability) said to the whole family that H.D. does not exist out here, is very rare, there's only a couple of families in the east who have H.D. An article had just appeared in the L.A. Times mentioning about 1000,000 Americans suffering from H.D. The article had mentioned the worldwide reach of H.D. From my experience with the average M.D., I know he is ignorant.

Question 4 - The problems and needs of H.D. patients. To prevail on them to keep an interest in living and family, to get along with friends and family, exercise, take enough nourishment, to cheer and bolster their spirits and try to make them understand that nothing is hopeless; there is work being done in research, which may result any day in an improvement, possibly a cure, not an easy task.

I have answered the questions on family and patients, as I have seen it through my experiences.

Question 5 - I have listed some of the problems these two children of my daughter, June Clark, have encountered in their short lives. I am with them, and will continue to help while I can, but I am 82 years old, and June becomes more difficult daily to care for, so the time is approaching when she will have to have residential care, and these two young people (I repeat, 17½ and 19 years of age) will have the overwhelming task of paying for the care of their mother, and also trying to make something constructive out of their own lives. The family estate has become very depleted through her long illness. There has been no genetic counseling, nor have I heard of any, only advice they have ever had from all the doctors was not to have any children.

I would be willing to testify, if necessary. Remember, I am 82 years old.

Mother of Professor Don Bradford and June Clark.
Granmother of Judy Rini, Dulcy Rimac, Drew Bradford,
Alan Clark, Amy Clark.
Great-grandmother of Melora Rini, Christine Rini,
Nicloe Rimac, Peter Rimac.

Los Angeles, California

April 15, 1977

DELLA E. BRASS
SAN DIMAS, CALIFORNIA

MARCH 15, 1977 .

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

BEVERLY BREIER
MANHATTAN, CALIFORNIA

MARCH 17, 1977

I am writing in regards to an acquaintance of mine, who has Huntington's disease. Her name is Ruth Krueger. I feel that something should be done on a nationwide basis. Perhaps the Government could help financially. A rest home would be most beneficial, as their condition tends to become worse with the passing of time.

She has tried to get financial help through the Social Security Disability program but was refused.

Hope you can do something for her and the hundreds of more like her.

Los Angeles, California

April 15, 1977

JOHN W. BREIER
MANHATTAN BEACH, CALIFORNIA

MARCH 17, 1977

I am writing in regards to an acquaintance of mine who has Huntington's disease. Her name is Ruth Krueger. I feel that something should be done on a nationwide basis. Perhaps the Government could help financially. A rest home would be most beneficial, as their condition tends to become worse with the passing of time.

She has tried, unsuccessfully, to get financial help through the Social Security Disability Program.

Hope that you can do something for her and the hundreds of more like her.

Los Angeles, California

April 15, 1977

TERESA E. BREIER
HERMOSA BEACH, CALIFORNIA

MARCH 17, 1977

I am writing in regards to an acquaintance of mine who has Huntington's disease. Her name is Ruth Krueger. I feel that something should be done on a nationwide basis. Perhaps the Government could help financially. A rest home would be most beneficial, as their condition tends to become worse with the passing of time.

She has tried, unsuccessfully, to get financial help through the Social Security Disability program.

Hope that you can do something for her and the hundreds of more like her.

KATHERINE M. BRIGGS
NORWALK, CALIFORNIA

FEBRUARY 14, 1977

Some of the problems that have been a major frustration to me deal mainly with convalescent hospitals. Aides and other help have a lack of awareness as to what Huntington's is and what care is needed.

You are forced to accept a convalescent hospital that will accept you. After an 11-month stay, a Bird Haven asked me to make other arrangements for Mother. The conditions in the nursing home she is currently in are appalling.

Most nursing homes are too large for Huntington's chorea patients who tend to get confused and wander. Most nursing homes are understaffed.

Now that she is unable to feed herself, she is fed by aides that literally stuff the food down with tablespoons so they can get on to the next patient.

She is now confined to a wheelchair when she is awake and restrained to her bed when not. She cannot communicate to say, "I have to go to the bathroom." Would anyone listen if she could? Now that she is confined to a wheelchair, she can't even get a drink at the water fountain. It does seem odd that a convalescent hospital would not have fountains at a lower level for wheelchairs. She has a great deal of difficulty drinking from a glass on her own.

Food is most often dry, instead of soft and moist to aid patients who have difficulty in swallowing. She has not quite reached the stage when all foods will have to be pureed.

Locating a doctor was another frustration that even CCHD was unable to help with. I was finally forced to accept one of the "house doctors."

There is a real lack of communication between the aides to the head nurse and the head nurse to the doctor.

I, myself, try to go in to feed Mom her midday meal, which is the main one, as often as I can. At these times I also observe the quality of care that she gets. The facility is small, I believe 60 beds, and at times is run smoothly and at other times is not. I have overheard on occasion that some

Los Angeles, California

April 15, 1977

days there are only four aides on duty. There doesn't seem to be any relief personnel available on days when help gets sick.

I hope this will be of some help.

Los Angeles, California

April 15, 1977

DENNIS BURGESS
BERKELEY, CALIFORNIA

MARCH 27, 1977

In September 1977, I met Jeannie Bjorklund when I was seeking to rent a room in her home. I will never forget that Friday evening: As I climbed a steep hill to Jeannie's house I saw the figure of a man appearing and disappearing from before the bay window. I knocked on the front door and Dr. Bjorklund invited me into the living room. Across the room I could see Jeannie lying on the couch watching television. Immediately her hand shot out in a waving motion. I waved back, but then her leg shot out in a similar action. I knew then that something was not quite right.

Jeannie was my first experience with a Huntington's victim. During the subsequent six months of her life I helped turn her over, walk her to the bathroom, feed her and pat her to sleep. And as I walk up that steep hill to Jeannie's home I still expect to see Dr. Bjorklund walking back and forth from the kitchen to Jeannie's bed caring for her.

Commissioners, I hope, and I say this most emphatically, I hope I shall never again see as much anguish as I saw on Jeannie's face during her life. Thank you for this consideration.

Los Angeles, California

April 15, 1977

HELEN N. BURGESS
STANFORD, CALIFORNIA

MARCH 18, 1977

Last year a friend of ours died in a veterans hospital. He was a victim of Huntington's disease. He had spent many years in the hospital and had hoped that through tests done on him some light would be shed on this dread disease. He had married and had children before he was aware that H.D. was in his family. He has three wonderful children. His daughter chose to have children even though she knew there was a 50 percent chance that she might become a victim of this dread disease. Her three daughters are exceptionally beautiful and extremely bright. She, of course, hopes that research will bring forth a cure for this disease soon.

I have seen the suffering the victim of H.D. went through before he died. He was almost completely isolated from the world through no fault of his own. His wife suffered and still suffers, worrying that perhaps one of the three children or one of the three grandchildren (should their mother fall victim to H.D.) might have inherited H.D.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

ANN BUSCH
FRESNO, CALIFORNIA

MARCH 21, 1977

I am a relative of a family who has Huntington's disease. I am not at-risk, but I'd like to add my name to your list of concerned people.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

OLAN BUSCH
FRESNO, CALIFORNIA

MARCH 22, 1977

I am a friend of a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

MARILYN CAMIN
FRESNO, CALIFORNIA

MARCH 22, 1977

I am friends with a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

KRIEG CAMPBELL
IDYLLWILD, CALIFORNIA

NOVEMBER 5, 1976

I am writing to you my deepest concern and most positive mandate to try to obtain Federal Funds for research into, mediation of, and eradication of Huntington's Disease and its deplorable ramifications. Time and effort not spent has no greater cost: our loved friends.

The tearful despiriting humility of trying to share oneself with afflicted friends and of trying to share the burden of potentially affected friends, is never forgettable. We need to improve our ability to diagnose Huntington's Disease. Federal funding to support all phases of HD research is desperately needed. Families having afflicted members need to be supported through home nursing care. We also need more people trained in all phases of HD patient care.

Commissioners, I am lucky. I don't have HD. I am also awkwardly lucky enough to have been involved with the afflicted.

We all know the tragedy it brings.

Commissioners, we must have a comprehensive National Plan. We need it NOW! It is a gloriously rewarded and absolutely necessary endeavor.

Los Angeles, California

April 15, 1977

S. KRIEG CAMPBELL
IDYLLWILD, CALIFORNIA

MARCH 15, 1977

I recently wrote to you of my concern about and support for research with Huntington's disease. I am very saddened to report the death of that afflicted loved one.

Commissioners--Go to Congress and President Carter on my behalf. It's no longer a matter of personal urgency for me; but for the thousands who have it and their children who have a 50 percent chance of having it, it is a matter of the gravest urgency. We are talking about cruel suffering, pain and death. Gentlemen, we can afford to fund.

Research, patient care and improved techniques of diagnosis must be given immediate priority and funding. I most strongly urge you to gather all power you can to conquer this horrible killer.

Call on me for assistance.

Los Angeles, California

April 15, 1977

BETTY CHAMBERS
SYLMAR, CALIFORNIA

MARCH 16, 1977

I have H.D. My smiles and happy attitude indicates that I am a woman and believe in God and Jesus.

This does not blind me to the fact of being misdiagnosis wrong medication for two years of sleep -- three of four children ran away to Alaska heavy with guilt. My fourth one never called or came over and always expects the worst.

All four feel better when someone stays with me. Although I personally love independence.

I can't clean or cook anymore. Two things I prided myself in doing before.

My three children invited me to Alaska. I was there three times--summer, autumn, winter (at Christmas) and spring.

Each visit was filled with love love and more love. My fourth child has come to grips with it. He is 31. My three in Alaska are 28, 24, and 22.

I go to church twice on Sunday. Relief Society twice a week, helping with baby nursery once a week. Go visiting teaching to five families a month. Go to the temple once every two weeks. Go to lectures once a week, watch bowling, eat out, go to dances to watch, make phone calls, work election board--crochet four (4) granny afghans for my kids. I learned to crochet two years ago.

It's wonderful to be alive at this time when H.D. Commission can do so much to help my loving family.

My daughter cries herself to sleep with worry that she will get it. My sons won't discuss it.

Thank you.

No one wants to get married and I am dying for a grandchild. We need help--any kind--every kind.

Los Angeles, California

April 15, 1977

DESTA F. CLARK
UNION CITY, CALIFORNIA

MARCH 2, 1977

As a relative of a Huntington's disease victim, I strongly urge your Commission to make every effort they can to research and cure it.

Los Angeles, California

April 15, 1977

EVA L. CLARK
UNION CITY, CALIFORNIA

MARCH 2, 1977

As a victim of Huntington's disease, I strongly urge your Commission to make every effort they can to research and find a cure for it.

Los Angeles, California

April 15, 1977

GEORGE CLARK
OAKLAND, CALIFORNIA

MARCH 13, 1977

Having seen the great suffering that Huntington's disease can inflict on its victims and their families, may I urge you and your Commission to make every effort to combat and find a cure for it?

Los Angeles, California

April 15, 1977

MAXINE A. CLARK
LINDSAY, CALIFORNIA

NOVEMBER 26, 1976

Mine is not an H.D. family. However, we have observed the heartbreak and tragedy that can befall these families. We have had close association with H.D.

So much needs to be done to educate our doctors of this terrible disease. My husband is a doctor and Chief of Staff of a small hospital. To his knowledge a case of H.D. has never been diagnosed there. Is this because our doctors would not know a case if they had one?

We are most interested in all aspects of the control of H.D.

Los Angeles, California

April 15, 1977

B. J. CLARKE
OAKLAND, CALIFORNIA

MARCH 13, 1977

In view of the enormous suffering that Huntington's disease can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

E. MICHELLE COOPER
LAWNDALE, CALIFORNIA

FEBRUARY 25, 1977

I am more than glad that a Commission has been formed to study H.D. I know this information came to me late, but I hope my letter will be helpful and will be counted. My brothers and I learned of our mother's disease in 1969. Our father passed away from cancer in 1956, knowing something was wrong with mother, but not knowing what. We spent what family money there was and all that could be spared on hospital, clinics, neurologists, G.P.'s, other specialists trying to learn what was wrong and watching her gradually lose control in walking and talking. She was scared, and sat on her feet to keep them from swinging. When she answered the telephone many unaware people thought she had been drinking. Mother never drank in her life. Finally, she developed pneumonia and my youngest brother, a lieutenant in the Air Force who was living with her at the time, put her in the Riverside Air Force Base Hospital. Dr. Greenberg, neurology, diagnosed H.D. the first week she was there. From 1956-1969. Thirteen years and finally we knew. We knew and knew not only did it affect our mother, but each of us and possibly our children. Also, we knew there was no cure and it appeared there was no research being done. No grants being granted to those who were trying. The public is mainly in ignorance that this disease exists.

We cared for our valiant mother until she could no longer sit up. I am a widow and work as an Eligibility Worker II for L.A. County. My eldest brother and his wife both teach school in Torrance. My youngest brother is still in the Air Force stationed in Denver. Mother was taken by "Bay Shore Sanitarium" in Hermosa Beach. She is still there. Dr. Greenberg told me he knew of no cases living longer than fifteen years with this terrible disease, but mother has. She no longer can move or speak but she is still breathing.

Finally, after we expensively and painfully learned about H.D., our eldest aunt told us their father had had "something like what she has." The family had been in darkness and held my grandfather's behavior as insanity. They were ashamed. On my grandfather's death certificate, cause of death reads "pneumonia," not H.D. and pneumonia. If only we had known. If only the public could be enlightened. I could go on with "ifs."

Los Angeles, California

April 15, 1977

Perhaps now you and your Committee will be able to accomplish what has never been done before. My hopes and sincere prayers go with you.

April 15, 1977

RHEBA CUNNINGHAM
TORRANCE, CALIFORNIA

MARCH 10, 1977

I am writing this letter with deep concern for my mother, Ruth Krueger.

Fifteen years ago, my mother had the uneasy feeling that she, too, was the victim of the same disease her mother had passed away with...Huntington's disease.

Unable to accept such heartbreak, she put the thought out of her mind.

Seven years ago my mother had gone to the doctor. Unfortunately, she was right. She did have Huntington's disease. She had quit driving that day.

That was in August of 1971. That December (1971) my father had a severe heart attack which laid him up for months.

While my father was recovering, my mother applied for Social Security. Since she has had a Social Security card since 1937 and worked up until 1971, she thought this would answer some of her prayers and also help my father get back on his feet.

To this day, my mother has been refused Social Security three times! They say she is \$50.00 or one point short. I think this is the most disgusting thing I've ever heard.

My parents had to sell their home and move to a mobile park so they could make ends meet. She deserves her Social Security payments.

One of the times after my mom had been denied Social Security, one rude employee from the Social Security office turned to my mom and said, "You can always try welfare!"

I don't think anyone should be treated in that manner. They have tossed her around like a ball, and it makes me sick.

I don't know how you can help, if at all, but I would appreciate a letter from you letting us know what can be done about this.

MARY DAVIDO
LOS ANGELES, CALIFORNIA

FEBRUARY 15, 1977

My name is Mary Davido; I am 28 years old, white, married, female. My occupation is at present word processing operator.

In reference to the psycho-social problems associated with the impact of H.D. on family members, I was around six years of age when my mother exhibited noticeable (to me) symptoms of the disease, eleven when she was committed (to the state hospital due to my father's inability to afford private nursing home care), and thirteen when she died.

I must assume a more or less "normal" childhood until the age of five and one-half; then my mother's condition deteriorated over the next five years. I missed at least one-quarter of each school year during this time; she didn't want me out of her sight. I was not allowed to go out in our yard and play, let alone have any friends. I had to cook for her, clean house, even comb her hair (if I pulled her hair during the combing, I was severely beaten). The first time she beat me, I told my father when he returned home from work and he wasn't upset, he laughed and said I must have done something to deserve a spanking. When I informed him it wasn't a spanking, but a beating with a stick, he did get upset and they had a big fight. The next day she beat me more severely and told me to never again dare tell my father about it. I didn't, consequently she did just about whatever she wanted to me during the next few years and my father never knew about it (one time she got carried away and blacked one of my eyes, and when my father came home he asked both of us about it. She said I was playing and hurt myself; I backed up her story. My father is not to this day aware of the brutality of her treatment of me. After she beat me (almost every day) she would be filled with remorse and try to cuddle me and baby-talk me, but I noticed that her remorse never stopped the rages or the beatings. She was getting worse, her speech was deteriorating, she fell down a lot, flew into violent rages over nothing, and had me totally terrorized. I had stolen a can opener and spoon and would creep around the canned goods shelves in the basement so she couldn't hear me, and open a can of food and eat it standing up in the basement. Had she found out, she would have beaten me for that too. This was because she, for reasons of her own, didn't allow me to eat in her presence.

I endured the years, and when she was committed, my relief was untold. My father was advised by a psychiatrist he finally forced her to go to that she must be committed before she killed me. He then asked me to tell him what she had done to me over the years, but I felt sorry for him, so I underplayed it, lied about it, and glossed it over. He then made the mistake of thinking that I had been too controlled by her, and I deserved some freedom, so he let me run wild. This was alright until I was twelve; then I discovered boys. I ran away when I was fifteen, and lived with a man who treated me just like my mother had. He flew into uncontrollable rages, beat me severely, and was filled with remorse afterward. I stayed with him five years, the same time period between my mother's deterioration and subsequent commitment.

During the time I was with this man, I had one child. I was seventeen then; when I was nineteen I requested and subsequently received a tubal ligation which was paid for by the state because of the H.D. I was counseled prior to the tubal ligation and was told it didn't have to be that way, did I realize that my mind might change when I was older, was I sure I wanted this? I was adamant; the operation was performed. I regretted having the one child (which was given out for adoption; I told the man I lived with it died), I wanted no more. I was unwilling to be a mother because of my fear of becoming like my mother. I would have died before putting another child through the hell my mother put me through. Also, I went through hell deciding not to have children, not to marry (I am not legally married to my present "husband;" we have discussed it and I tell him if I notice myself exhibiting my mother's symptoms, I will leave him), not to get involved with a man I couldn't leave if I developed the disease. I was not about to face someone else with decisions like this.

My present husband was informed of my condition while we were dating. He thinks I am wrong; not about having children, but about leaving him if I develop the disease. I watched my father's life ruined by my mother--if he hadn't loved her, he might have been able to leave her before it got too bad, but since he did love her, he tried to convince himself that it wasn't there. He will never remarry; he says that he realizes there isn't one chance in a billion of anything like that happening again, but...he's still not going to chance it. I don't really blame him; a lot happened prior to her commitment that I have purposely left out. One other thing I would like

to mention is that after my mother was committed, she wrote to people she knew in high places (local government) stating that her husband had railroaded her into the institution, there was nothing wrong with her, etc. A lot of the people she wrote to went to the institution to visit her--one picture was worth a thousand words--and that was the end of that; except for her best friend next door, who believed her and to this day vilifies my father.

In conclusion, I believe this testimony speaks for itself regarding some of the problems caused by H.D. In speaking of the period after I ran away (including to great extent the present), I underwent quite a few experiences most people would rather avoid, in the name of "Live now, who knows what the morrow brings?" because I was very aware of H.D. hovering over my horizons; if I was to develop the disease in my life I wanted to be able to have experienced "everything," to have lived life to the fullest before it happened. This was also what caused me to have that one child--I wanted the experience of birth, even if I could never allow myself the experience of being a mother. I have always worried about dying and regretting all the things I didn't have a chance to do; so I've probably done as much in my 28 years as most people do in a lifetime.

I hope this is sufficient. Thank you.

Los Angeles, California

April 15, 1976

RALPH A. DEAN
LOS ANGELES, CALIFORNIA

FEBRUARY 10, 1977

My acquaintance with H.D. has been through knowing a group personally who has lost one member of the family and has another member who is afflicted.

There are three children, adults, in the family who are possible victims. Two of these children each have two of their own who are exposed to H.D. Several of these small children were conceived after their parents knew of the presence of H.D. in their family.

My first reaction to this seeming indifference on the part of the parents was to the end of instituting massive educational programs for all people who are subject to H.D. to convince them that the only way at present to help stop H.D. is to not conceive children by parents who are subject to H.D. It is very wrong to bring children into the world who may develop it. Even though some children may not develop it, they may pass it on to their own offspring. It should be mandatory at the time of applying for a marriage license for both parties to state whether there is or has been H.D. in their background.

Research programs should be initiated to enable clinical examination of any individual to show if he is subject to H.D.

An educational program should be developed to convince H.D. people that they should submit to sterilization until a definite cure has been found.

Anyone who has observed the trauma that occurs in families of H.D. victims will understand the need for radical preventive methods.

DORIS DE SOUZA
SACRAMENTO, CALIFORNIA

MARCH 15, 1977

I am an "at risk" person of Huntington's disease. I am 50 years old, but I don't feel at all free of getting the disease. In my own family we have it start as early as 27 and as late as the 60's. There is quite a group of us. All of my grandfather's brothers and sisters had H.D. and six out of nine of my aunts and uncles were afflicted. Each one has his own story.

I grew up with H.D., but not knowing its name. I knew something was there but I wasn't going to let it happen to my children. I was going to the doctor and do something about it. So I had five children. Every time I would hear a name put to our problem, I'd look it up. When a doctor from Stanford said Huntington's chorea and I looked it up and every symptom was the same. I saw six doctors. (I thought it was just my family--thought I was all alone. Since then, we've found there are thousands, so we are definitely not alone.)

One doctor told me it was so rare that if he saw another H.D. patient in his lifetime, he would be lucky. One doctor told me I knew more about it than he did. My own doctor asked me to write all I knew about it for him. There is a great need for the doctors to know there is research being done. There is also a great need for a place for H.D. patients where the attendants will know how to take care of them and just because they can't understand what the H.D. patient is trying to say, they are by no means retarded. The mind of the H.D. patient is very, very alert, long after they have no control over the rest of their body. It is just a long, hard, heartbreaking life. I know there is no pain, but to say it is distressing is too mild. It's ugly to say the least, and for much too long we have had no hope.

This started for me when I was a little girl. A little girl came to play and she said, "What's the matter with your grandfather?" I said, "There's nothing the matter with my grandfather." "Well, why does he shake?" I answered, "He's old, that's why he shakes. All old people shake." My grandfather was 54 when he died, at least eight or ten years after that conversation. He was in an asylum in Norman, Oklahoma. So was one of my uncles.

Two of Mom's brothers loved horses and visited us at fair time every year. They could not dial the phone so they started walking home. The police always thought they were drunk until they talked to them. They would always bring them home.

Until recently, we didn't know enough to plan ahead for H.D. So no one had adequate insurance and most insurances, I understand, have a no neurological disease clause. None of my family has been financially able to take care of extra patients. We have only been able to put them in the care of the county. Even if we could pay, there is still the problem of attendants not knowing how to handle or understand H.D. patients. I heard one lady say she put her mother in a home and later when she went to visit, the mother was in a straight jacket, yelling and making all kinds of fuss. The attendant found her walking outside. She had these violent motions and not talking clearly, so they put her in a straight jacket. That was the easiest for them, I suppose. The daughter said, "Sure, she's mad. She's mad at you for putting her there. If you just listen to her, she's cussing you out for putting her there!"

We were lucky with my mother. She got H.D. sort of late and it was slow. She was 68 when she died, two years ago.

I'm helping to start a C.C.H.D. satellite group in Sacramento. Our group wants to give information and comfort to anyone who contacts us. Your group could help by getting the medical field educated to H.D. and a place for terminal H.D. patients.

Los Angeles, California

April 15, 1977

DORIS DeSOUZA
SACRAMENTO, CALIFORNIA

MARCH 26, 1977

I am an "at risk" H.D. person. I wrote a letter to the Commission but did not say I would testify if needed. My concern is the great need for doctors to be educated about how to diagnose H.D. patients and to know all the research that is going on; and the need for convalescence homes for H.D. patients with attendants who know how to treat the patient.

Los Angeles, California

April 15, 1977

MRS. D. DiMERCURIO
MOUNTAIN VIEW, CALIFORNIA

MARCH 12, 1977

Out of eight brothers and sisters, seven have this dread disease. Three have already died, two are in the last stages and another two are showing signs of having it. In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

Three children of the above mentioned have the disease also. God knows how many in the family will have it.

Please help!

Los Angeles, California

April 15, 1977

MRS. H. DiMERCURIO
SAN JOSE, CALIFORNIA

MARCH 11, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

MARY DiMERCURIO
MOUNTAIN VIEW, CALIFORNIA

NOVEMBER 5, 1976

I am writing to you because my husband has Huntington's chorea. In the newsletter you asked that we write and tell of our problems with it.

Well, to begin, my husband has had it for about 18 years. The last five have been the worst, of course. I can't remember how many times I've had to rush him to Emergency at the hospital with some injury because he has fallen. I've been sure he would put his eyes out one of these days. His entire body is covered with bruises because of accidents.

I couldn't continuously follow him around. Now for the last four years he has been bedridden, although keeping him in bed is really hard; he keeps throwing himself out of it. I have to feed him, wash, dress, shave, and put him up and down on the commode.

I am really at the end of my rope. When he first went to a doctor about H.D., they didn't even know what it was. I have three daughters and with my luck, they will all get it. It's terrible for the person that has to care for a loved one like that too. And horrible to think they have no chance at all. I think family counseling would be a great help and also some kind of therapy, of course. After all these years, you might say we've about given up hope.

In my husband's family of six sons and two daughters, everyone but two so far have it. You can see why I've lost all hope.

Please, please do something to help research this horrible disease that has caught so many unaware. I could write you a book about my experiences with Huntington's chorea but I'm sure you will get many letters like mine, perhaps more coherent than mine and plainer. So that's about it.

Los Angeles, California

April 15, 1977

MARY DiMERCURIO
MOUNTAIN VIEW, CALIFORNIA

MARCH 10, 1977

My husband is a victim of Huntington's disease. I wish you could just take one look at him, and I know you would do something to combat it.

I can't tell you everything that he has gone through and also my children and I who are waiting to see who it's going to hit next. What torture it is to live with the thought constantly on your mind that you are going to be just like that. I have taken care of my husband at home.

He has had it since 1960 and there is nothing to him at all, but skin and bones. Now I wonder if I have done the right thing, but it's too late.

I can't tell you in this letter what hell we've been through. I would need a book for that.

Please, please, I urge your Commission to do something to help combat it.

HELP!

Los Angeles, California

April 15, 1977

TOM DiMERCURIO
MOUNTAIN VIEW, CALIFORNIA

MARCH 10, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

KAREN DREBING
FRESNO, CALIFORNIA

MARCH 22, 1977

I am friends with a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

JOLEN E. DUNWOODY
MILPITAS, CALIFORNIA

MARCH 10, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

MARY E. DUNWOODY
MILPATIS, CALIFORNIA

MARCH 10, 1977

In view of the enormous suffering Huntington's disease can inflict on families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

JOAN EDWARDS
FRESNO, CALIFORNIA

MARCH 22, 1977

I am a friend of a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

JO ANN EKHARDT
SAN FRANCISCO, CALIFORNIA

MARCH 26, 1977

I was diagnosed as having Huntington's disease in September 1972, at U.C. Hospital, when I was 49 years old. At the time I was heartbroken and terrified, having witnessed the ravages of the disease on my father, who died when he was 70 in a veterans hospital. He had been a tremendously vital and talented man--an artist, James E. Allen.

The reason I went for a diagnosis was because I was having difficulty performing my job at the Internal Revenue Service. The condition, then largely mental, had become progressively worse, and I had trouble accomplishing the daily necessities.

At that time, I had worked for 24 years, and should have been in a position to receive a livable annuity. However, I had worked for ten years for private industry (mostly as a draftsman) before starting work for the Government, and due to recent changes in the law, am no longer eligible for Social Security benefits without working five more years.

Also I had once borrowed from my Government Retirement Fund, and unless I repaid \$2,874.00 immediately, I would receive only \$210 a month.

I finally decided to apply for Disability Retirement, after much deliberation, and received it in March 1973.

Then I managed to work for two years for private industry, and repaid a bank and credit union loan, so that I paid back the money due my Retirement Fund, and received the higher amount of \$320 a month. I was let go from my last job in February 1975 and haven't worked since.

Due to inflation, I'm getting worse off financially by the day. My rent has already increased by \$70.00 and is due to go up again with the raise in property taxes and doubling of water. I have had to spend nearly \$1,000 on dental work, and now starting this year, my disability annuity is taxable. I find this unbelievable--to add hardship to hardship. Over the years my annuity has increased to \$430.00 but this is not a commensurate sum for 26 years of labor.

I would like to keep working to reinstate my Social Security, but I feel so inadequate. My memory is so undependable and my mind confused. I never know when I'm going to forget the most basic information, and must keep copious notes. This is very embarrassing, and noticeable to some fellow workers who don't understand and aren't sympathetic. It puts one under a constant strain.

Although the loss of one's mental faculties is unpleasant enough, I realize things can get much worse. My father was reduced to violent shaking fits almost continuously, and had to be hospitalized for his last ten years.

While I am still able to halfway cope, I'd like to be able to provide for a livable annuity and future hospitalization. I was married twice, but both are dead, and I have no children.

Now I have a number of physical symptoms. My walk is erratic, and I often lose my balance. Almost daily I notice physical deterioration one might attribute to malnutrition. I was taking Haldol every day as prescribed by my physician. This resulted, I think, in the appearance of hundreds of congealed blood spots under my skin. I stopped taking any medication, except for Synthroid, but in the last few months, my nails have become ridged, my veins have become prominent and I've developed brown spots on my face and hands. I don't know what to eat to prevent these abnormalities.

I feel H.D. is certainly a dread and fatal disease that leads to years of utmost disability and despair. There is little hope but for a cure. If this ever becomes a reality, it will be people such as Marjorie Guthrie who made it possible.

Los Angeles, California

April 15, 1977

NANCY ELLIOTT
DALY CITY, CALIFORNIA

APRIL 4, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future generations, I STRONGLY URGE your Commission to make every effort to support research and assist in every way to combat and cure it!

Los Angeles, California

April 15, 1977

RUTH ENGEL
TURLOCK, CALIFORNIA

MARCH 28, 1977

I have Huntington's disease in my family. I lost my mother and older sister and an older brother from it and a younger sister and a younger brother who are in a nursing home with the disease.

Anything you can do to help to find a cure or even a drug or operation to arrest this disease would be an answer to my prayers.

I am 60 years old and have for thirty years worried about getting it.

In view of the enormous suffering Huntington's can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

ALBERT ESPINOZA
FRESNO, CALIFORNIA

MARCH 22, 1977

I am a friend of a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

SAMMIE EVERSON
FULLERTON, CALIFORNIA

MARCH 25, 1977

A PERSONAL EXPERIENCE

A skinny thirteen-year-old girl was helping me make cheese sandwiches, which we were selling at a benefit concert in Topanga Canyon. It was hot and the cheese was melting but people were buying them like hotcakes. We decided to take a break and listen to the concert for awhile.

Lisa and I walked through crowds holding hands as we found an empty bale of hay. I sat on the hay and she sat in front of me.

Will Geer was reciting a poem by Robert Frost. David Carradine was singing one of Woody Guthrie's songs. Everybody was singing along and having a good time. Toward the end of the concert Will Geer told the people why they were there: to contribute to the fight of Huntington's disease. He was explaining some of the symptoms as well as other aspects of this horrible disease. How Woody Guthrie suffered and died from H.D.

As he was speaking, Lisa clenched my hand and said nothing. I knew what she was feeling. She turned and looked at me with tears in her eyes and questions in her face.

Her mother has H.D. as did her grandfather--Woody Guthrie. Her questions? Do I have H.D.? How do I plan my life? Should I have children? Should I even get married and put someone else through this horror of not knowing? Chance it to pass this thing called H.D. on to my children? When will I know? I may not have H.D.!! I will know when I am 30-40 years old. When I am 30 or 40 years old will I then sit in a hospital bed with uncontrollable movements like my mother? Or will I have a full and happy life? How can I? With all these questions, will somebody care enough to find some answers for me?! So I can plan some kind of a life for myself? Please??

Lisa is now 16 years old and in love. Still no answers. Still nothing but questions. As so many people have that have that 50-50 chance of having H.D.

Somebody has got to care enough to give these people something to hope for!

Research is a start and it is the only way; it's a start but it isn't enough. Not now! We need more doctors that understand the disease and can diagnose the disease. Hospitals that will accept H.D. patients. Education of medical personnel to deal with H.D. patients. I worked in a convalescent hospital as a physical therapy assistant, where Lisa's mother was a patient. When she first came to our hospital no one knew anything about H.D. We soon learned. Mary Guthrie Boyle taught myself as well as the staff of our hospital all she could about H.D. That's how I got involved with the fight for control of H.D. We started working with Gwen and tried to walk with her. It was hard at first, but we did work with her and after a few weeks she was walking with the assistance of one person.

As we kept working with her, at times her movements would quiet down a lot and she would feel more at ease with her surroundings. She was even enjoying some of the activities in our hospital.

That was two years ago. She is now in another hospital and in very bad condition--but just because H.D. is a progressive disease doesn't mean we have to sit back and let it take its course! We have to give them some hope--show them that we care! Through physical therapy or other therapy--just show them that someone cares! Please don't take away all their hope! Through P.T., I know that we can maintain some control. For some time at least--but it is some hope! The only answer, of course, is research. Please give these people a ray of sunshine--not just for themselves but for their kids and grandchildren.

Thank you for listening to me.

Los Angeles, California

April 15, 1977

CHRIS FLORES
FRESNO, CALIFORNIA

MARCH 22, 1977

I am a friend of a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

BRENDA FOGATO
STRATHMORE, CALIFORNIA

MARCH 22, 1977

My father was a victim of Huntington's disease. I am at risk and therefore feel very strongly that Federal funding is needed.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

NEVA FREUDENBERG
INGLEWOOD, CALIFORNIA

MARCH 27, 1977

I have known Ruth Krueger and her family for over 20 years. Her mother was a victim of Huntington's disease. At that time they did not know what she had, until years later. Then one by one the three children came down with it. Ruth's older sister, Betty, younger brother, Don, and then Ruth.

The family was in a state of shock, not knowing how to deal with it.

Ruth worked at the bank for a few years before she knew the disease was in the family. She had to quit because of the disease, and is unable to collect Social Security, which puts quite a hardship on her. She worries about the financial burden it will bring as to the care she will need, when she can not mobile on her own. There is no aid open to her.

Any help she can get would be greatly appreciated.

JESUS REYNOSO GARCIA
SAN FRANCISCO, CALIFORNIA

MARCH 14, 1977

Being the youngest (33) of ten children born to a Huntington's carrying mother, I know intimately the pain and suffering this devastating disease can bring. My mother was already sick when I was born. My father, in his ignorance and desperation, was having her treated, in vain, by a doctor who knew nothing of the symptoms, much less the cause or cure for such a puzzling illness in the early forties.

I never knew a "normal" mother. I did know, as only a sensitive child could, that something was wrong when people called her "crazy" and she shook uncontrollably and unexplainably at times. God, how she must have suffered in that world of her own. Most of the time she was kept at home, hidden from the outside world, a prisoner of that deteriorating mind.

Occasionally she and I would go to visit my godmother, but it was I, a little six-year-old boy, who would take her by the hand and carefully lead her across the highway.

You can't begin to realize what it was like, at eleven, to be separated from a mother to whom I had been so close, incarcerated in that state asylum.

And the worst agony, for me, was having to visit her and listening to the cries of anguish and sorrow behind those locked steel doors. I shall never forget the sadness in that once beautiful face as she tried to explain something about having been abused and stripped of her dignity. I guess, in a sense, it lives on in me.

When Melquiadez Reynoso Garcia died in 1957, she left a void in my life that can never be filled. I could never seriously think of getting married and having children with even the remotest possibility that they might become victims.

I was thirteen. She died of pneumonia they said, then they thought it might be Parkinson's disease, but after an autopsy, it was diagnosed as Huntington's.

My eldest brother, Leonard, died in 1973, also a Huntington's victim, leaving behind a successful business, a young wife and five at-risk children.

Josephine, my eldest sister, broke down at Leonard's funeral and was diagnosed the next day. She is in her late forties.

It has skipped my second eldest brother and sister evidently, but a younger brother and sister, in their early forties, show all the obvious symptoms.

For the rest it is really too soon to tell, but it is not a rosy future to look forward to.

And, for myself, I really don't know--but I don't think a day has gone by since I was fifteen, when I realized what I might be carrying, that I haven't thought about it and wondered. The memory of my mother being so clear.

I remember once, several years ago, when Mrs. Guthrie was in San Francisco, organizing a Committee to Combat Huntington's Disease Chapter, suggesting a slogan:

Huntington's--another word for HOPE.

Meaning that by publicizing this destructive disease and making people aware of the need for money or Government grants for research of Huntington's and other genetic related diseases, only then could they begin to hope for a control, for a cure, and possibly for a little peace of mind.

Gentlemen, are you going to deny us that hope?

Los Angeles, California

April 15, 1977

URSALA M. GETTLE
SAN JOSE, CALIFORNIA

MARCH 10, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

SALLY ANN GILLESPIE
WEST SACTO, CALIFORNIA

MARCH 27, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

April 15, 1977

BARBARA GIORDANO
CARMEL, CALIFORNIA

MARCH 20, 1977

Personally, I haven't any idea of what to say in this letter. The problems of being at risk and having a mother who has had H.D. for 11 years seem obvious to me. I am, of course, willing to do anything I can to help, so I am writing this letter.

I am 15. I have a younger sister and two older brothers. We are all at risk. I have had to grow up very quickly and have become much more responsible than others of my age group. It was especially hard when I was younger because I didn't understand that my mother was "sick." I have suffered the same pains as any child who has lost a mother.

Nevertheless, I am very close to my mother, although it isn't at all a mother-daughter relationship. Now I can talk with her as a friend. But when she was at home the roles were reversed. I was the mother and she was the baby. It was very confusing (my mother is now in a nursing home).

As for being at risk, well, I can't worry about it. If it happens, it happens. I'm not going to ruin the life I have by worrying about it. It used to bother me, but now I ignore it.

Also, my friends never pressed me with questions. Everyone has problems!

I can tell you one thing, I am not going to have children! If I marry and want some, I'll adopt. H.D. has to be stopped if it can't be cured.

Well, I have candidly stated some of the facts. I hope they will help. If I can do anything else, please let me know.

Los Angeles, California

April 15, 1977

MARIA GIORDANO
CARMEL, CALIFORNIA

MARCH 20, 1977

I am at risk to get H.D. I am at the young age of 13-1/2 years old. My mom has Huntington's for about 13 years. I believe everything should be done to wipe out this disease by the time I am 21.

Because my mother has had this disease almost all of my life I have some very strong views on this subject: one is that all people should know about it.

My father is having a fund-raiser on June 11. Our family friends, Joe Conley and Eric Scott from the TV show, "The Waltons," are going to come up for it.

April 15, 1977

GREG GIORDANO
BELMONT, CALIFORNIA

MARCH 8, 1977

I am writing this letter partly at the request of my father and partly upon my own initiative, to relate what life has been like for an at-risk male individual with a Huntington's disease stricken mother.

I am a twenty-year-old male, honor college student in my junior year at the College of Notre Dame in Belmont, California. My mother is presently stricken with H.D. and is in a convalescent hospital for medical care. My father is a college professor at a junior college in Monterey, California. My brother, Jim, is presently a sophomore at San Diego State University. My two sisters, Barbara and Maria, are 15 and 13, respectively. The older one is a sophomore in high school and the younger is in eighth grade and both are presently living with my dad in our home in Carmel, California. I hope to attend graduate school for a career in law or as a CPA. The only thing holding me back a little is my financial situation due to the added expenses of my mom's illness and the rising prices of both hospital care and higher education.

The area which is hardest hit by my mother's illness is logically the family and home life. Fortunately for me and my brother, my mom was stricken after we had pretty much grown up and had the benefit of an upbringing with two healthy parents. Unfortunately, I can't say the same for my two sisters, especially Maria who was born just a few months before my mom became noticeably ill. She thus didn't receive all the personal care and motherly love so necessary to a growing child; especially to a female. My father couldn't supply all the special things which a mother supplies to a growing girl.

The strains this disease has put on our family have been enormous. I'm quite proud of the way our family has held together and weathered this storm. My dad has had to not only do the normal duties of a father but also take on all the duties of a mother. Gradually as we children got older, we all accepted a share of my mother's normal duties, including a few of my father's too, so as to relieve some of the strain. Thus at early ages we were all cooking dinner, cleaning the house, gardening, doing the laundry, shopping,

driving around town on family errands, etc.; all the things which would normally be done by a healthy mother. These added duties brought responsibility and maturity to all of us at a rather early age. But this was not done without a few costs.

For one, my father has had an obvious curtailment put on his social life with a sick wife and has had to make many sacrifices of his own time to take of other family members. Too, we kids have been hesitant about having guests over to avoid embarrassing my mother with a situation in which she couldn't control herself. Third, I'm sure that all the added pressures and responsibilities have taken a few years off of my dad's life (strain on the heart, etc.).

Another problem I found as I grew older was what responsibility did I have during dating and later in marriage, as an at-risk individual? Should I risk it and have children of my own, or play it safe and not have any of my own? I strongly feel that the best thing research could find right now, besides an out-and-out cure, is a method of detecting early whether one will or will not get H.D. This development would be very important for future at-risk individuals who would like to have a family of some kind and yet not endanger any offspring.

In closing, I cannot help but stress the hardships and costs which this disease has put on our family and many others. I would like to hope that we can count on our Government's support through your Commission in assisting us in our fight to lead normal and fulfilling lives while faced with the grim possibility of contracting this disease sometime in the near future. H.D. not only strikes the immediate victim but also hurts the rest of the family as well. Your support and assistance is greatly needed and appreciated.

T. JAMES GIORDANO
MONTEREY, CALIFORNIA

MARCH 28, 1977

I'm writing in regard to an ailment that has plagued my family for the past nine years. This catastrophic disease I'm referring to is Huntington's chorea. Our family has had the menacing and cruel burden of suffering through our mother's ailment for as far back as I can remember. My mother, who I feel has been awfully brave through the disintegration process, has also suffered extraordinary pain.

My mother, Jackueline Giordano, not only feels her own personal pain from the day-by-day penetration of the disease, but feels the added dimension of knowing her children have a 50-percent chance of contracting her same dreaded disease.

Many times I've laid awake at night, unable to fall asleep, thinking of what I would do if I contracted the same tormenting disease my mother has. I've contemplated whether I would immediately join the army, to die heroically in battle, or to submit to the same ugly and lonely years of agony as my mother has. For I can honestly say my mother is very lonely. All her old good friends seem to have slowly deserted her when she's needed them most. I know how it must hurt.

I also anticipated the possible and puzzling question of what I would do if I, with the luck of God, failed to get Huntington's chorea but one or all of my three sisters and brothers got it. I dread this situation more than my own inheritance of the disease.

I just hope if I am faced with any of these possible outcomes I will be blessed with the same courage, perseverance and faith my father has displayed in coping with our situation at home.

Our astounding financial burdens as a result of the numerous medical expenses are immeasurably outweighed by our family's constant tension, pain and anguish.

My personal career goal is to become a doctor, so I can possibly make a direct attempt at conquering and curing the inflictions caused by Huntington's chorea. I'm currently a junior at San Diego State University, majoring in both biology

and physical education. So if I can keep the good grades and the proper frame of mind, and with the guidance of God, I just may achieve such a goal.

Let's don't wait until my lifetime to donate money and appropriate funds to fight Huntington's disease. Let's do it now so we can save my mother and all those like her from submitting to a horrible, inevitable outcome.

I have very few faint memories of when my mother was well and active. But I can honestly say these were probably the happiest memories of my life. I just wish my younger sisters could share in these same memories. For my sisters have always known pain and can only think of my mother as sick, sick, sick... I love my mother and I want to help her any way I can. I want my friends, my enemies and the whole damn country to help her. She damn well deserves it!

NADINE B. HAITH
ARLETA, CALIFORNIA

DECEMBER 29, 1976

Huntington's disease was diagnosed through an autopsy when my husband's brother died in September, 1976. While we had known about and suspected H.D. for four years, his family would not accept the fact that it could be, and therefore did not get him to a doctor for help. He had chorea and the last five years he also had mental deterioration. If he left the apartment, he would get lost and couldn't find his way back. While his wife had never worked outside the home, the last three years she was certainly needed at home to look after him. We first noted his lack of coordination and feel he had H.D. all of 15 years if not more. At the age of 54, he was not able to work any longer and had to go on disability. At that time the doctors said he was senile as a man of 70. He died at the age of 62.

My husband's sister, age 45, has had a mental problem for 13 years and the doctors have felt it was because she had so much trouble in the delivery of her son. In November 1976 she was given tests at the University Hospital in Omaha, Nebraska and was diagnosed as an H.D. victim. The doctor there has treated 24 H.D. patients, so she at least is now getting the right care and not shock treatments that they previously had been giving her.

A brother died in 1971 at the age of 49 in a mental institution, and it took my husband six months to get a death certificate, cause of death "too much smoking." At the time we were not aware of the H.D. in the family, but as we look back, we feel this very well could have been his problem.

At the present we have another brother, age 48, living in our home who has been an alcoholic for the past five years. His wife is divorcing him, he just had no place to go, and could not make any decisions. His children refuse to accept the possibility of H.D. and feel he is an alcoholic. My husband has contacted his family doctor who had heard of H.D. but doesn't know anything about it. However, he is concerned, has sent for the material from the National Committee, and has set up an appointment with a neurologist for him. He may or may not be a victim of H.D. If he is a victim and is not able to work, what is to happen to him?

April 15, 1977

Just knowing H.D. is in the family has been a strain on all the family members as we all have grandchildren that we may have passed it on to. I am a grandmother, age 58, married, and work as an office manager in a high school of the Los Angeles Unified School District. My husband is 56 and as yet is not a victim of H.D.

Los Angeles, California

April 15, 1977

SALLY J. HAMMOND
SANTA CRUZ, CALIFORNIA

FEBRUARY 26, 1977

As a friend of a family which is afflicted with Huntington's Disease, I ask you to do everything possible to improve treatment, facilities, and research and training in order to fight this insidious disease.

In the family to which I refer, it is the mother who is afflicted, and it is extremely distressing to know that her four healthy children have a 50% chance of getting the disease--and of passing it on to their children if they are afflicted.

I have seen their mother, who is now in a convalescent hospital (even though she has, at this time, no hope of recuperating), and I have felt total helplessness. It is for this reason that I write this letter.

In view of the enormous suffering Huntington's Disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

FREDERICK G. HARRIS
WALNUT CREEK, CALIFORNIA

MARCH 13, 1977

In view of the enormous suffering Huntington's Disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

ARLIE HATFIELD
WATERFORD, CALIFORNIA

FEBRUARY 7, 1977

My wife is a victim of Huntington's chorea. This is truly a most heartbreaking disease. One has to go through it to really know what all the problems are. There are no more times for sharing things or talking together or companionship. There is no rest and no time to take a break. It is a 24-hour babysitting job, with the agony of knowing it only ends with the death of your loved ones. Hardly none of the doctors we consulted knew anything of this disease. I think all doctors, as well as the public, should be more knowledgeable about this disease. Family counseling should be there whenever you need it. There are times when you need to talk to people who know or have been through the same as you.

I would like to see rest homes or convalescent hospitals built for H.D. victims only, so they can be cared for properly. I also think research for this disease should be greatly supported. Not only is it unknown and unfamiliar, but many have it and it's very deadly and heartbreaking.

I hope my words touch the hearts and minds of some of you for your support.

Los Angeles, California

April 15, 1977

MARLENE H. HENRY
PALO ALTO, CALIFORNIA

APRIL 5, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

EUGENE HIKEL
SAN DIEGO, CALIFORNIA

APRIL 1, 1977

My wife, Sherry, has H.D. in her family. Her father died of it and her sister just recently died of it. For several years we have suspected that Sherry has it, however, we cannot get any doctors to diagnose her. She shows many of the symptoms that her sister did. We have taken her to several doctors but none confirm our belief. This has been a big problem. Her sister had H.D., but it was diagnosed as a spoiled brat. It was not until she had a child that they diagnosed her as having H.D. Then I think it was because her husband was in the Marines and they wanted to know what was wrong with her. Unfortunately the medical profession is hesitant to or won't diagnose the disease.

Another major problem I will be facing is possible hospitalization of my wife. This will be a major expense and I do not believe I will get much assistance. I am in the middle class and as a consequence we receive little assistance for major problems from either the Federal or State Government. I believe this is totally wrong.

A third problem is the inability of diagnosing H.D. early so that people can make plans about their families. It would be good if a means of early diagnosis could be discovered.

Finally I hope a cure is found soon.

Los Angeles, California

April 15, 1977

SHERRY HIKE
SAN DIEGO, CALIFORNIA

APRIL 1, 1977

My name before I married was Craig which was a descendant of the Rathbuns, which there is a genetic study of. My sister, Cathy, got H.D. when she was 16. We took her to a neurosurgeon. He said she was a spoiled brat. Doctors do not give a diagnosis of having H.D. I have been given tests for the past five years and have been given the diagnosis that I did not have H.D. I know that I have. Having to see a beloved one have H.D. is the hardest thing there is.

Los Angeles, California

April 15, 1977

LEONA K. HILLEARY
RICHMOND, CALIFORNIA

MARCH 24, 1977

All you can do to seek out a way to combat Huntington's disease will never bring my father and sister back, but you will have my eternal and everlasting thanks and prayers for your efforts toward that end.

Los Angeles, California

April 15, 1977

RHONDA HOUK
STRATHMORE, CALIFORNIA

MARCH 22, 1977

My father was a victim of Huntington's disease. I am at risk and therefore feel very strongly that Federal funding is needed.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

INIS HOWARD
VENTURA, CALIFORNIA

MAY 20, 1977

My father died in 1969. He had Huntington's disease. He died of a collapsed lung and other complications. His sister (my aunt) is in a rest home--she has Huntington's disease. Although it wasn't diagnosed as such, their mother (my grandmother) surely must have had it. She died years ago in a home for the mentally ill. Looking back, my father had some of the same symptoms as my grandmother, although I was quite young when my grandmother died.

This puts me in the "at risk" category. I am now 44 years old. I am married; have three children. We knew there was something wrong with our dad several years ago, but thought it was related to Parkinson's disease. It wasn't until my aunt went to doctors to see about getting treatment for Parkinson's that we learned it was Huntington's disease that she had; then we knew that was what Dad had, too.

Most of my family lives in Missouri and we only see them on vacations, etc. I have been able to keep the threat of Huntington's disease from my mind only because I wasn't around any of it--"forget it and it will go away," you hope. I haven't even been terribly familiar with all the effects it causes. However, I do remember clearly my dad getting weaker as the years went by, his speech getting more garbled each year at vacation time. Then, finally, him being completely bedfast, completely dependent upon my mother to care for him, feed him; unable to go to the bathroom. I have four sisters and two brothers, so there are several of us "at risk." One sister has it already, we are sure.

My husband and I went to Missouri right before Christmas this past year, and another sister went to see a doctor about herself. I sat in the doctor's office with her as he confirmed her suspicions--she definitely has the beginning symptoms of Huntington's disease. I can't describe to you the awful, sick, desperate feeling I had at that moment. She seemed to be accepting it just fine. I think she had suspected (as we all have through the years) that she had it. But I was completely knocked off guard, because I hadn't been around her for a while, and she seemed "normal" to me. She is 4 years older than I. After the first initial shock of learning that she

has it--then it hit me like a ton of bricks!! I probably have it too, and it will begin showing up soon. Most of what I had read and heard on the subject led me to believe that if you made it through your thirties, then you were pretty safe. Here I had made it to 44 years old and was feeling pretty good, like I had it made--and then came the realization that if it just started showing up in my sister at age 49, then more than likely it would start showing up on me in the next few years or so. I didn't have it made after all--I PROBABLY HAVE HUNTINGTON'S DISEASE!!

Now the daily wondering and agony--every time I'm a bit nervous, or dropping things, or forgetting things, or accidentally stumbling, or not being able to type as well as I could; it's a relentless watching and waiting, feeling every day that I surely must have it!! And, of course, this makes me more nervous and jumpy.

My brothers and sisters and I have always joked about it when we saw each other--we would look each other over good and say, "Have you got it yet?" or "You look okay to me," etc. Now it's no joking matter, since my sister has actually been verified as having Huntington's. Another sister hasn't been to see a doctor, to my knowledge, but she has had these symptoms for several years, the same ones I can remember my dad starting out with; and we feel that she has Huntington's disease.

So, now, with this newer feeling of being "at risk" comes the horror of knowing that if I have it, then one or two or all three of my children will be going through the same agony and horror as I, because they'll be "at risk," also. They will have to wonder if or when they will get this awful disease. They don't remember their grandfather too clearly, but I do. One of my daughters is already married and so far has no children. I was feeling pretty good, looking forward to my first grandchild--but now with this threat over me (it has been there all along, only it's just intensified since I found out my older sister has the disease and that makes me a prime target right now) I don't know what to advise them to do about having children. That's a decision they will have to make between them. Then my 16-year-old daughter will have to make the same decisions when she marries. My 7-year-old son will have a bit longer before he has to decide anything like that.

I can recall my dad going to town with us sometimes, and people thinking he was drunk, the way he was staggering around, and his speech all garbled. His head bobbed up and down and around, and he made funny faces, moving his mouth around a lot. His mind stayed alert, seemingly. That was another agony for him and my mother--he could understand her but she couldn't understand what he was trying to tell her. The last few years, especially his last year, he would just sit or lie in bed and the tears would flow down his cheeks as he watched us and he couldn't say a word to communicate. He would try--making grunting noises, but the words just wouldn't come out. We used to make a game of it with him--like playing "twenty questions."

I remember the kitchen linoleum being terribly worn out under the place where my dad sat to eat his meals, when he was still able to be up. His feet shuffled around so much that he had the floor worn bare, practically. He knocked things over easily at the table.

I am hopeful that some day there will be a cure or something to halt the effects of Huntington's disease. I hope and pray my children won't have to watch me through the years, each year getting progressively worse, as I had to watch my dad. I also hope this Commission will take this letter to heart--as well as the many other letters that have surely been received--and will request Federal support for the research into the cause of Huntington's, and in turn will be able to find a cure to end this dreadful disease!

Los Angeles, California

April 15, 1977

MRS. CRETA MAE HUNT
FRESNO, CALIFORNIA

MARCH 25, 1977

We need your "Help!"

Our daughter is adopted. She is 21. Her real father died in his 40's of Huntington's. She also has a sister who started showing signs of having Huntington's around the age of 12, now she is 24 and in a rest home. We believed until just recently that our daughter wasn't going to get Huntington's. Now she is pregnant and the doctor tells us that he sees signs of Huntington's and they are running tests now. As you know, if she has Huntington's the baby has a 50-50 chance of getting it. We as a family who knows the heartache and suffering that a family and the patients go through, ask for your help, for all of us who have been through this live on hope that someday through research there will be a cure.

We have been to visit our daughter's sister many times and to see a young girl at the age of 24 in a rest home. It breaks your heart, that there isn't a special hospital or a wing of a hospital with trained people to help the ones with Huntington's. The care and treatment they need instead of letting them sit in a chair and die. I don't blame the nurses and doctors at the rest homes because how can they treat them when they don't even know what Huntington's disease is? I believe that if there was a special hospital or place for patients with Huntington's, that it would be a help in research for a cure for Huntington's. We pray with all our heart that our daughter doesn't have Huntington's, but the doctor doesn't give us much encouragement.

When we get to the point where we feel like what can one person do and then our Government tells us we are all important, for us to speak up and be heard, and they will listen. I know what I am speaking about. I have lived with Huntington's disease and I have seen the suffering. SO PLEASE HELP US! in our research for a cure and help for all Huntington's patients and families.

From a mother who needs to know there are people who care and that there is hope.

Los Angeles, California

April 15, 1977

OSCAR L. HUNT
FRESNO, CALIFORNIA

MARCH 24, 1977

Please help us. My mother died at the age of 43 from Huntington's disease after five years of being bedfast.

There are eight of us children in our family and five of them have Huntington's disease.

I had a brother, Charles, who died at the age of 47 from H.D. Another brother, Lloyd, is 34 and will soon be bedfast.

Two of my sisters, Emma and Mary, are in rest homes at the ages of 38 and 40; another sister, Cora, at the age of 36 has H.D. but her husband and three children are still trying to make a go of it.

If you could have seen what I have seen in my 42 years of life!

To try and feed your mother, brothers and sisters and still see them slowly starve to death because they cannot control their muscles to swallow and to think of the children they have that will be at risk in having H.D., you would not hesitate to pass any bill that would give all the people that have or will get Huntington's disease a little hope that some day there may be a cure and care for the patients that need it now.

Thank you.

Los Angeles. California

April 15, 1977

MRS. JACK HURLEY
TORRANCE, CALIFORNIA

MARCH 16, 1977

I am writing this letter in the hopes it will give some help to my neighbor, Mrs. Ruth Krueger, a victim of Huntington's disease.

She does not receive any Social Security and we feel she will soon be in need of a rest home. She is a lovely person and never seems to complain, and we feel she is very deserving of any help available.

Los Angeles, California

April 15, 1977

EVELYN M. ICKES
TORRANCE, CALIFORNIA

MARCH 17, 1977

I am writing in regard to an acquaintance of mine, who has Huntington's disease. Her name is Ruth Krueger. I feel that something should be done on a nationwide basis. Perhaps the Government could help financially. A rest home would be most beneficial as their condition tends to become worse with the passing of time.

She has tried, unsuccessfully, to get financial help through the Social Security disability program.

Hope that you can do something for her and the hundreds of more like her.

Los Angeles, California

April 15, 1977

JAMES F. ICKES
TORRANCE, CALIFORNIA

MARCH 17, 1977

I am writing in regard to an acquaintance of mine, who has Huntington's disease. Her name is Ruth Krueger. I feel that something should be done on a nationwide basis. Perhaps the government could help financially. A rest home would be most beneficial as their condition tends to become worse with the passing of time.

She has tried to get financial help through the Social Security disability program but was refused.

Hope that you can do something for her and the hundreds of more like her.

Los Angeles, California

April 15, 1977

JULIE JACOBSEN
FRESNO, CALIFORNIA

MARCH 22, 1977

I am friends with a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

April 15, 1977

IDELL JACKSON
VILLA PARK, CALIFORNIA

MARCH 4, 1977

As a concerned citizen who has been caring for an H.D. patient for the past year, I would like to make a brief statement about my experience with the illness and its tragic consequences.

First of all, the illness is so rare that few doctors would be qualified to recognize the symptoms and diagnose it properly, thus delaying help and resulting in financial loss to the family involved. Secondly, as the illness progresses and brain loss ensues, the symptoms become very bizarre. To wit:

My "patient" has lost control of that part of the brain which dictates chewing. She is compulsive about stuffing food into her mouth and swallowing it whole, resulting in constant fear of choking and suffocation. She now has no sense of time and if she is looking forward to something that will take place the next day, she may get up in the middle of the night and insist upon being bathed and dressed for that event; all the while crying and pleading and making life quite miserable for all those around her. These temper tantrums increase with the progress of the illness. Also, she may get up and go out in the night (to get the morning paper, check the mail box, etc.) giving constant concern about her falling and harming herself.

Since their inability to walk decreases as time goes along, there is the constant concern over their falling, doing themselves serious injury. This, of course, necessitates more and more confinement, restricting their own lives and that of their families. Also, the emotional trauma of all this and its effect on the husband and children is very sad indeed. Vital young persons who are at-risk for this disease must watch all of this, knowing that they may be a rerun of the whole episode; knowing also that if they marry or have children they will be subjecting all of them to this eventually. All the ramifications of the illness would take some time to discuss, but since it is a genetically inherited disease it certainly must be considered one of the major health problems of our time.

Los Angeles, California

April 15, 1977

It is certainly hoped that something can be done soon to help alleviate the suffering of victims of H.D. as well as their families.

April 15, 1977

HELEN L. JEFFERSON
NEWBURY PARK, CALIFORNIA

APRIL 6, 1977

No words can begin to adequately describe the various agonies, frustrations, financial problems and losses, emotional involvements, etc. caused by Mr. Jefferson's illness.

He and I have been married almost 38 years, and during most of that time we have faced problems related to the disease. A very few years after our marriage, we began seeking help from a variety of specialists for Stan's nervous problems. Even after the facial grimaces and beginnings of the choreiform movements began, no suggestion was made that he might have a medical problem involving the central nervous system.

Mr. Jefferson's employers began changing him from job to job, and though he was a very intelligent and talented young man, he seemed to have problems expressing himself. In spite of that he was given heavy responsibilities, but then the day came when he began to be demoted because of his hesitancy in speech and other problems involving his judgment.

Our marriage was under tremendous strain, and by this time our teenage daughters were faced with strange reactions and one daughter actually married against my wishes and good judgment because her father sanctioned the marriage and became belligerent with me because I opposed it. The groom was even worse than I predicted and in a short while she was left with two babies and \$7,000.00 (about) of debts and a husband in jail for the second time. She divorced him because he was impossible. I KNOW she wouldn't have married this young man, had her father reasoned with her--we had always been consistent in our training of the children up to this point and had a very close relationship as a family.

Fourteen years ago Mr. Jefferson's health had deteriorated to the point that he was placed in a hospital for two weeks for an evaluation, and he was released with a "clean bill of health" even though he was so spastic in his movements he could barely get his fork or spoon to his mouth!

At that point he ran away from home, lost all the money we had had in a home we had sold before his last move and a

brand new car which was completely paid for. He was going to buy a tractor and haul for North American Van Lines. In six months after starting that venture, he wrecked the truck (not his fault, but had not paid the insurance and ended up penniless and half-starved at the Salvation Army in Chicago). They kept him for six months (he refused to come home) and gave him psychologic and physical help. Finally, in answer to our prayers, he consented to come home; and at that time at last specialists in the Los Angeles area diagnosed his condition as Huntington's disease and told me of his hopeless condition.

Now I was faced with the problem of HIS care, managing somehow to see that our younger daughter finished her training as a nurse (she did receive her B.S. and the bills were paid), helping with the expenses of the older daughter's family and paying off the debts with which she was left, and working full time! As the result I suffered physically and have been under the doctor's constant care for many years. Yet, there is no other way for me but to continue working full time. This September I will reach 62 years of age, and feel I should be able to retire, but there is no way I can continue to pay for Stan's institutional care and live myself on Mr. Jefferson's retirement and Social Security incomes. It takes almost all of that for his care and medicines and insurance!

I am a secretary, have a serious hearing impairment and one rather blurry eye! I am under constant strain endeavoring to maintain top performance in my job.

In addition to all of the above, I might add that Mr. Jefferson was a minister who was highly respected and admired for his many talents, in spite of his slowly progressive impairments. Because of his action in running away after being told he was physically fit, the influence of his work was undermined. We were cut off almost entirely for several years socially. I was even told by one former friend, "I can't be seen with you. I will phone you and we will still keep in touch." This was almost more than one human could endure, but this is par for the course when one's mind is affected as tragically as Mr. Jefferson's.

1. At this point in time I work full time.

2. I maintain a home I've managed to buy, but still have a mortgage on and owe more than we paid for a home just before it was sold and the equity of which he lost.

3. I bring Mr. Jefferson home weekends because he does love to be home and becomes extremely depressed if I do not. He requires special foods because of his swallowing problems, so this means a lot of extra work weekends for me.

4. We cannot have a normal social life--a few of our old and true friends do understand and can watch him eat if I invite them for a meal, but with food falling out of his mouth, and the contortions he goes through to eat, it is revolting.

5. He will soon be unable to walk, yet we face years of slow deterioration. Because he lived such a clean and healthful life, physically he is still in good condition, except for the Huntington's--I mean organically he has no problems.

6. This disease has affected almost all of our 38 years of marriage. Financially it has placed a tremendous burden on me. Yet there is no help available, unless I am reduced to the poverty level. I could desert him, but we loved each other too much for me ever to do that, and our children are so devoted to him, none of us could stand such an "out."

7. We daily face the realization that one or both of our daughters may suffer the same tragedy in their lives.

8. We daily face the reality that our four grandchildren may also suffer. We cannot anticipate the future with anything but dread. We can live day by day and live as normally as possible, but we all know what can and probably will happen.

9. Our plight is less tragic than many because I have been able to work and because Mr. Jefferson is covered by Social Security and our church's retirement benefits.

10. We have a faith in God that sustains, but many families are not as well informed of Bible concepts.

11. The restrictions placed upon me by these circumstances are less traumatic for me because I love gardening, reading, studying, painting and other art outlets and am contented at home. However, I do become very lonely and long for family support and cannot afford to see our daughters as often as I would normally. I have tried to shield the children from seeing their father often because it tears them apart--yet they are loyal to him too.

12. Our 15-year-old beautiful Kathe Michelle said one day, "Oh, Gommie, I wish Gompie were well. I can't even remember him different from what he is now."

This is only a thumbnail sketch. I really do plan to write a book on Stan's life and how we have all reacted to this horrible illness and its ramifications. I haven't even mentioned many areas that will take a chapter or two! And we still haven't been able to trace the illness in his mother's family because of her early death and her mother's early death from TB.

Anything you can do for Huntington's disease patients is needed and will be appreciated by those whose lives have been shattered. The hereditary factor is the monster that terrifies.

April 15, 1977

ROXANNE JEROME
ESCALON, CALIFORNIA

MARCH 3, 1977

I, myself, have been exposed to a family of dear friends who are victims of suffering Huntington's disease. Something must be done to help all we can to act on this spooky and suffering disease. I am not much of a writer nor a speller but I must put my point across to you all in behalf of what I have seen and felt for my best friend and her family.

Their mother is a victim of H.D. and is now in a home being taken care of. I've never visited this place but just from her daughter's words, it's a losing battle.

The daughter, Margo, one of four daughters, made a most painful and in my eyes sad decision. She sterilized herself from fear of ever bearing a child who could inherit H.D. Margo is 24 years old and quite a strong and unselfish person to have made this choice. A wise choice, yes, but unfair to her womanly desires to be a mother.

There must be something we all can do to avoid having people we care and love so much from experiencing such an ordeal of great fear. I do not know much about H.D. myself, but I do know plenty about how it has altered my girlfriend's life. Please, I urge your Commission to make every effort to combat and cure this God awful disease. Please, please help! I am willing to do anything I can to help Margo and her family plus the many other families afflicted with Huntington's disease. Will you please be willing to help us?

Please, let's all join together to help our brothers and sisters to live.

Los Angeles, California

April 15, 1977

DAVID L. JOHNSON, M.D.
SAN RAFAEL, CALIFORNIA

MAY 19, 1977

I am a 46-year-old former physician. For two years I have been unable to practice because of Huntington's disease. Memory loss was my major problem, and I have a tremor. Last year I had to stop driving because of Huntington's disease.

My major reason for writing is my 11-year-old son. I would like to see vast amounts spent in research on the cause of Huntington's disease in hope to prevent it before he has my miseries.

I would also like work done in finding a good test for Huntington's disease so that I don't have to worry my son unnecessarily about this horrible disease.

April 15, 1977

MELODIE JOHNSON
SALINAS, CALIFORNIA

MARCH 28, 1977

My name is Melodie Johnson and I am writing this letter for my mom.

My mom is Verdis Johnson, and she has H.D. After showing some symptoms of H.D. she had the painful brain scan and some other tests run on her. We found out that she had H.D. in October 1972. I was in the sixth grade then. The doctor that handled her case put it very bluntly that she was going to get worse instead of better.

My mother and father didn't like the way the doctor handled her case because he was so blunt and he had never dealt with a person and their family with H.D. so they switched to another doctor. Dr. Hack is her doctor now; he is a neurologist and a psychiatrist. He had heard of H.D. and had done some research on it but had never dealt with a person that had it. The medicine that Dr. Hack prescribed for my mom is Etrafon 4/25 #150 and Ritalin 10 mg. #200. This medicine has helped her a lot and the doctor seems to think that she is getting better instead of worse.

Out of seven children in Mom's family, five have H.D. My uncle, Berlin West, who lives in Alabama, has H.D. and is in the Tuscaloosa Hospital. He doesn't recognize his wife and other relatives when they come to see him any more. My aunt, Juanita Green, who also lives in Alabama, has H.D. and she forgets a lot. Aunt Burma Phillips and Aunt Lyniss Burnett also have it and they, too, are in the same condition. H.D. struck them in their late 30's but it hit my mom in her late 40's.

When my grandfather died, the doctors had not really heard of H.D. They said that he had died from a brain tumor. My uncle, Jerry West, did some researching though, and found out that he had died of H.D.

I'm adopted so I can't get the disease, but my sister is 28 and she might have it and her little boy could have it when he grows up. Hopefully, with everyone's help, H.D. will be taken care of before he is old enough to have it or before his children are.

MARIAN JOHNSTON
TRACY, CALIFORNIA

JANUARY, 1977

The ignorance among doctors is something I have encountered many times. Recently I had to go in for surgery. I explained to the doctor that I had Huntington's disease, therefore I didn't know how my muscles would react under anesthesia. Later he told me that he had searched everywhere he could think of for information about H.D. He said it was impossible for him to find anything. Information was not available!

I spent many years trying to figure out what was wrong with me. My local gynecologist finally sent me to a neurologist in Modesto, California. He diagnosed it as H.D. Before the diagnosis was finally made, I had gone to several different doctors and their attitude was that it was "all in my head." Imagine my frustration when I believed that there was really something terribly wrong and no one would acknowledge it. Even my family and friends wouldn't believe there was anything medically wrong.

When I was a child in a small town, my father had had a car accident and was unconscious for several days. After that, doctors told my mom that the cause of his uncoordination was because he had injured his head in the accident. The opinion of the townspeople was that he was a drunk. He was even jailed because of his uncoordinated walk and slurred speech.

The family (seven children) had to go on state aid because he couldn't work. He had been a sought after carpenter. One of my high school teachers saw him and got my mom to take him to the University of California Hospital at San Francisco. It was so long ago that I was not aware what the diagnosis was.

I was the first in my family to be diagnosed with H.D. Family patterns indicated that both my father and my older brother also had the disease. My father died by being hit by a car. My brother got to where he couldn't take care of himself and ended his own life with sleeping pills.

The neurologist in Modesto, California asked me if any of my children had children. Then he told my husband and I to tell them not to have any children. One daughter was pregnant at the time, so I didn't tell her that I had H.D. until after her delivery. The impact on her was deep depression for over a

year. I have four children that are facing the future with Huntington's disease. I now have five grandchildren and one great-grandchild.

One aspect of myself is that it is almost impossible to write letters. A daughter is writing this for me.

I feel a real need for Federal funds for H.D. research. I am willing to help with research. If I can give my body and my time, won't you please give your money? For myself, my children and any others that face the terror of Huntington's disease.

I used to be a person everyone loved to be around. Now it's hard to find good things to talk about and share with people. Give me hope.

April 15, 1977

MARIAN C. JOHNSTON
TRACY, CALIFORNIA

MARCH 9, 1977

I am a Huntington's disease patient. My daughter is helping me write this letter. I am writing because I feel there is a need for more research on Huntington's disease. In this letter, I will relate my personal experiences as examples of problems that Huntington's disease patients encounter.

Personally, I have encountered much ignorance in doctors on the subject of Huntington's disease. Although I tried in vain to tell three different doctors that I knew that I had a physical problem, including loss of coordination, none recognized any problem, sending me away with the assurance that "it was all in my head." This after even attending a very formal dinner at which I met one of my husband's supervisors and I was unable to shake hands with him even though I tried! My coordination was bad and I could not control my hand.

The fourth doctor I approached, a neurologist, was finally capable of making a correct diagnosis. Imagine the emotional strain I felt when I was told it was "all in my head!" My own perceptions were being ignored.

Again, I encountered doctoral ignorance and even lack of availability of information when I later had to have an unrelated type of surgery. I informed the doctor that I had Huntington's disease and asked if and how it might cause difficulties during the operation. Not only had he not heard of the disease but he also could not find information on it when he searched and in asking fellow doctors.

Obviously, doctors should be better informed about Huntington's disease.

Another of my biggest concerns as an H.D. patient is over the future of my four children. I strongly feel that good counseling should be available for both patients and those who are at-risk. My own children have experienced great difficulty in adjusting to the high pressure of not knowing whether or not they will get Huntington's disease. One of my daughters went through a time period in which she lived only for the "fun" in

each day. With the help of a good counselor she is now able to think some in terms of her future.

H.D. patients are normally depressed. Without the help of an excellent psychiatrist and counselor, I would probably have ended up in the state hospital. Instead, I am living at home with my husband. I am able to function normally both at home and in my community, six years after having been diagnosed as an H.D. patient.

I feel fortunate to have been able to afford to go to the psychiatrist of my choice. But one of my big concerns about my future is that my husband is at retirement age, and after he retires we won't be able to afford the \$30.00 an hour two times a week that I paid for the psychiatrist. This may mean that I won't be able to afford the counselor of my choice as my problems become more severe.

My biggest worry is my children and my grandchildren. (I was already a grandmother when I realized I had H.D.) They need money for research. My two daughters and I are concerned enough about research that we flew down to Los Angeles to donate spinal fluid and blood and were tested neurologically for the Hereditary Disease Foundation tissue bank.

We are always available to help with research, but unfortunately research hasn't been in enough volume to use us often. We have only been called upon twice in the last six years. How I wish that there was so much research to help us that we were tested constantly in an effort to find a cure for H.D.!

Los Angeles, California

April 15, 1977

L. W. JORGENSEN
FRESNO, CALIFORNIA

MARCH 21, 1977

I am a very close friend of a family who has H.D.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

PATRICIA KAUFMAN
ATWATER, CALIFORNIA

MARCH 3, 1977

My Mother has Huntington's Disease. The trials and tribulations we have gone through for the past ten years are tremendous. It took us that long to get a proper diagnosis for my Mother. It was only through an autopsy performed on her brother that we were finally able to determine what caused the horribly weird behavior and deterioration of a once beautiful person. Prior to the autopsy, it was years of tests, hospitalization, psychiatrists, mental institution, shock treatments, etc for her. Had we known what was wrong with my Mother, we could have avoided so many tragedies which are too numerous to name.

Now we are faced with the problems of my Mother's care. She has an income of \$27 per month. We cannot get social security for her. Nursing homes don't want Huntington's patients, nor can we afford one for her. The only alternative is a mental institution. Huntington's disease patients do not belong in mental institutions.

After visiting my brother and his family last summer, I found we have an extra burden to bear. The doctors had tested him for four days and are 99% sure he is also afflicted. He is 42 years old and has three beautiful daughters. What do you tell the daughter, who had just been married, when she asked "Dare I have children"?

As for myself I try not to dwell on the fact that I am in the high-risk category, but each time I stumble or cannot recall a name, I cannot help but think "Am I next". I am 40 years old. What is in store for my four beautiful children? Will I end up being a terrible burden for my husband?

We urgently need your help to find a cure for Huntington's Disease!

Los Angeles, California

April 15, 1977

FLORA E. KELLY
TORRANCE, CALIFORNIA

MARCH 21, 1977

On hearing of the mid-April meeting to be held in Los Angeles, I wish to lend my voice in deep concern about my neighbor, Ruth Krueger, whom I have known close to three years.

Before she was stricken with this dreadful Huntington's disease, she worked and paid into Social Security for over twenty years. She tells me she came short of a mere \$50.00 in qualifying for disability through Social Security.

The H.D. alone is a big enough problem for one to cope and live with every day, without the addition of the financial impact. And this is exactly what happens--when one's employment is forced to come to a halt through no fault of their own.

In consideration for the disabled, it seems to me her being denied was cruel, when she had paid in her full amount, except short of \$50.00.

Another point I wish to mention is Ruth's dear Mother was also a victim of H.D. and before her passing, she was placed in a mental hospital, and she really wasn't mentally ill.

With so many thousands of families confronted with H.D. it seems it is time for government-paid rest homes.

I thank you for reading my letter, and do hope it will be given kind consideration.

I wish to add that I am a widow, age 61, and I work part-time as a sales lady.

Los Angeles, California

April 15, 1977

BETTY KENNEDY
CLOVIS, CALIFORNIA

MARCH 22, 1977

I am friends with a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

RUTH KING
TUOLUMNE, CALIFORNIA

APRIL 7, 1977

At last I am getting around to writing. I only hope I'm not too late to do any good.

Now, where to begin. It all started back when I was in the seventh and eighth grades. I can remember my mother and I were real close up until then. That was when she began to change and got sort of hard to live with. She always had an excuse about what was wrong with her. I mean, like when she couldn't walk a straight line anymore, it was because she had a planter's wart on the ball of her foot. Her speech being funny was because of her teeth needing work. She was good at always finding excuses so she wouldn't have to face the fact that she was a victim of H.D. I guess you can't blame a person for trying to block that part of their lives out of their minds. But it did make it hard on me, as I didn't know what was going on.

Through my high school years, it was like hell at home. I didn't dare disagree with my mother or talk back to her to any degree, or she would work herself up into a tantrum and believe me, that was terrible to watch, besides being scary. She threatened my life several times, beat me with a shoe one time, always was yelling at me, broke a yard stick over me, and on and on and on. It is too painful to go over and over. I ended up running away from home a week before my graduation from high school. But because of my father, went back home for awhile.

Then came marriage for me right after I graduated. So here my mother was, getting worse and worse, and all the time me getting to hate her more and more. Yet, she still was my mother, so I was always going into her home and doing things for her, like cleaning and such. Finally it got so bad that she was falling and calling anyone that would talk to her. She was going down to neighbors' houses, neighbors she didn't really know. I was starting to get phone calls from the people telling me to come get her because they were a bit afraid of her and she was making a pest of herself. Only now do I realize she was just lonely. My aunt, who without her help I would have never made the bumpy road ahead, and I decided it was time to get her to a doctor. That is a story in itself. What a laugh! I would go from doctor to doctor and explain the situation, and all I would

get from them is an offer to give me a prescription for her to calm her. Thanks but no thanks. We needed more than that. Finally I came across a doctor that was very interested in the case and was glad to take her on. He is still with her today. Thank God for doctors like him. I wish there were more of them.

Things were getting pretty bad with my mom, to a point where she was no longer to be trusted alone. Then came the battle with the county. What a headache! It took quite a long time to get a worker and do all that paper work. It took quite a bit of convincing to make them see we really needed their help because my dad in no way could pay the expense of a rest home. They had to come see her for themselves, and then I had to sign more papers and more papers. I know and see people on welfare that could be out here working, and yet when we who really need help go to get it, we sure get the static. It was a long, drawn-out thing to get help, and I wouldn't want to go through it again.

Then came the rest home. How do you think it feels to have to trick your own mother into one of those things? Not very good, I'll tell you. She wasn't able to control her body, but there was nothing wrong with her mind, so she didn't want to leave her home. After all, who does? That was a terrible day, I will never forget her pleas. But there was nothing else we could do. I tried to get her into a home with an excellent reputation, but they were all filled up, so I took the next best (?) one and put her on the waiting list at the good one. Well, in six months, she was almost dead. I was killing my own mother. She hated it there, and they treated her awful. That home wasn't fit for pigs. How places like that stay open is beyond me. It's cruel. My mother wasn't getting out of bed at all, for anything. So I went over to the home that had her on their waiting list and actually begged them to take her. They didn't have any room for her, but because of a kind and understanding man, he made room for her. For a home, it was really neat. It had a more homey feeling and she was treated good. The girls took time with her when she didn't get mad at them for something and start hitting at them. When she started swinging was when all her strength would come back. She could sure get strong.

She is now in the same home but they have expanded and built new buildings so it has lost a lot of its homey feeling, but compared to others, it is still better than most. She is just

there now. I don't know how much she understands and how much she doesn't. She is skin and bones, if there is any skin left. I don't know how much longer she can last, but then I said that three years ago too. She never has talked about the H.D. thing; she fought it all the way down.

My dad has survived, but barely. There was a time when I thought he was going to go under. The county took so much away from him (or the state); so much that he had worked real hard for much of his life. They allow him so much to live on, and then take the rest of his check, no matter how much he makes. Things have to be put in my name so we're sure when she goes, the state doesn't come and take what little he has. It took a while, but he has gotten used to having only so much freedom.

I am so grateful for the H.D. organization. I am going to the meetings in Modesto, as they are the closest meetings. For so many years I felt so alone in this thing. I thought I was the only one on earth with a mother such as I had. Then came the newspaper with the story about Mr. Guthrie. Wow, I wasn't so alone after all. I started writing letters like crazy. Finally I started getting answers. Was I ever glad.

I could go on and on and on. I have so much in me about all my experiences and ups and downs. There have been so many tears shed over the years, more than I dare say. This H.D. is a terrible and ugly thing, and hope to God that someone can help. The only thing that gets me up after being down, or if I start thinking sad, is that I do have some life free from H.D. if I'm even going to come close to getting it, is that I'm so better off than some people. I'm thankful for what I've got today and I will face tomorrow when it gets here. The only thing that really scares me is I don't want my family to have to go through what I had to go through. It was too ugly for too many years. I'd do anything to spare them. But like I said, I don't worry about it because I'm living for today.

I hope they are close to getting some help and assistance for people who have H.D. in their family. Why should it be such a headache to get a doctor and financial help if needed? It's not fair that people with a sore toe should get help easier than us. How can we get people to wake up? I'm glad there are people like you guys who want to fight. I'll help!

Los Angeles, California

April 15, 1977

Thanks for listening. If I can be of help just let me know. I can tell you a lot more if you are interested. There's all kinds of ugly things and hard things. In fact, I could write a book. But not now.

Los Angeles, California

April 15, 1977

E. A. KIRKPATRICK
PACIFICA, CALIFORNIA

MARCH 5, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

MARGERY G. KOEHLER
TIBURON, CALIFORNIA

FEBRUARY 28, 1977

I am Margery G. Koehler, one of six children of Peter and Nellie Borree born in Hayward, California 58 years ago. Our family, three sisters and two brothers grew up in the Hayward area where our father was born and most of the children lived during their early married years. About ten years ago, our brother, Richard, he was 52 at the time, had pains which he thought was arthritis because his father had suffered from this and Richard's doctor was treating him for arthritis. About this same time, spring of 1965, our mother had a malignant cancer removed from which she recovered and returned to her home where she lived alone after the death of our father in 1963. In her later years Mother had an uncontrollable motion in her legs which her doctors just passed off as a nervous condition. Mom's younger sister, Mrs. Alta Julianel, went with Mom on a visit to Mom's doctor at which time he diagnosed Mom's nervousness as Huntington's chorea; and this was the first time any of us had heard of this disease. However, the doctor was unable to give us any information regarding it. Up until this time our Mother had appeared to be a very healthy person and it was only at age 81 she moved to a convalescent home and lived out her last year there.

Mom would be heartsick if she had known the seriousness of this disease and this is passed on to her children. It is certain she had no knowledge regarding this at the time she was having her children. This is why I want to help; and with everyone working together we will find a cure for Huntington's chorea in my generation.

About four years ago our family learned our brother, Richard, had Huntington's chorea and began to realize the implications of what this meant to our family. After reading an article in a publication indicating what we might expect as time passed, I decided to become better informed and to become actively involved. I wrote to the Southern California Chapter which was newly formed asking what we might do to help and received a newsletter and solicitation for funds. We continue to receive the newsletter and contribute as much as possible; my husband and myself both have small businesses so our time and funds are somewhat limited.

My brother's condition was stabilized with the use of drugs and he was comfortable and able to maintain his home with his wife. He was able to go to the local market by bus on his own to buy groceries; we also took him shopping frequently and he was able to handle the grocery carts and make his own selections.

There is now a Northern California Chapter of the Committee to Combat Huntington's Disease. I don't know just when it was formed, in Oakland, California. From the time we heard of the Northern Chapter we have contributed to it in the hope that we would be able to get more help in regard to our brother's condition. There have been several meetings of this chapter but we have been unable to attend due to our business commitments. In the May 18, 1975 Newsletter from the Northern Chapter, it asked for people willing to help in a research project. Thinking this would be an opportunity to help, I filled out the application form for volunteers and took it to the Oakland Chapter headquarters. This was a large medical building with no listing of the H.D. Committee so I went to Suite 60A which was the address listed on the Newsletter. The doctor at this office was very rude and tried to give me the brush-off; finally, at my insistence, he referred me to a doctor on the seventh floor and I again received the same treatment. No one seemed to know about the H.D. Committee. This is where we had been sending contributions and so became concerned that the Northern Chapter was just a big rip-off. We went so far as contacting the Better Business Bureau in Oakland, but have never had a response from them.

I spoke to our family doctor in Mill Valley and showed him the Newsletter hoping that he might be able to help, but he was not aware of the H.D. Committee and possibly not even of H.D. All this occurred during July, 1975 and at this time my brother's wife fell and broke her hip and was taken to the hospital and my brother tried to get along by himself at their home. About two weeks later my brother had to be hospitalized; he was found in a coma and was failing rapidly when he arrived at the hospital. My sister, Mrs. Vivian Wentworth, and myself talked with the attending doctors and asked if they were treating Richard for Huntington's disease. When they said, "No," we insisted that they contact Dr. Zwange, neurologist, who had been treating Richard for many years. My personal feeling was that they thought Richard was under the influence of alcohol. Physically that is the impression Richard gave, but a doctor

should be able to tell from a few simple tests. In May, Richard had attended a family picnic and had then flown, alone, to Reno to visit their daughter and grandchildren. I talked with him when he returned and although his speech was slurred, and more so when he became excited, we were able to understand him quite easily. After several weeks in the hospital he and his wife were well enough to be transferred to a convalescent home in the Hayward area. Richard was now on stabilizing drugs at the request of Dr. Zwange, Valium, Thoroizine, and Artane, one of each three times a day.

We then learned that Richard would not recover, and I recalled reading about the development of a tissue bank for the victims of H.D. We then contacted the hospital in the L.A. area and were contacted back by Dr. Goodlett Woodworth at the Veterans Administration Wadsworth Hospital Center. Dr. Woodworth described to us in great detail and length what we would need to do and the procedures to get the sample tissue to them in proper condition in L.A. When Richard died, we followed the instructions and had excellent cooperation from the mortuary and hospital in getting the autopsy and sample properly packaged for air shipment.

Several months after this, we received a phone call from Dr. J. Harmand Bjorklund, President of the Northern Chapter, apologizing for our difficulty in locating them and giving us a phone number where we can contact him if need be.

ALVIN KOSLOFSKY
SAN JOSE, CALIFORNIA

DECEMBER 16, 1976

In 1974 my wife died of Huntington's Disease. I just read an article which stated that you will be holding a meeting in Los Angeles about mid-April of next year. The article encouraged victims of HD to share some of their personal experiences and problems with you in the hope that federal resources could be appropriated to help eradicate HD and help the families financially.

My wife had the disease for eight years before she died. During that time she slowly deteriorated, physically and mentally, to the point where she could no longer walk or talk, had no bladder or bowel control, had to be fed, and tied into her bed. The last two years she was in a convalescent hospital, I could no longer take care of her at home. During that time, our 3 children--the oldest was 12 at the time of her death, I needed to hire a combination baby sitter, housekeeper, and nurse's aid. Good ones are next to impossible to find. I had to pay up to \$500 a month for one; medicine and doctors--doctors had trouble diagnosing the illness and didn't know what to prescribe because there was no known cure, yet they tried different drugs, cost around \$300 a month; and the convalescent hospital charged \$700 a month. I had Blue Cross insurance which helped some. However, they didn't want to pay for the hospital because they called it a non-recuperative illness. So with about \$1,500 a month going out for the above reasons and needing to meet the necessary food, clothing, and shelter expenses for the rest of the family you can see what that does to a middle income family financially as well as emotionally.

The article referred to above quotes Senator Edward Kennedy as saying, "We're going to take your recommendations seriously...We cannot wave a magic wand and just by dollars alone find a solution to these diseases...But the U.S. has the extraordinary capacity to give a disease priority in terms of federal resources." I sincerely hope so.

I know there are no easy answers, but it seems to me that if our great country could spend \$150 billion on the war in Vietnam, or appropriate money to study the sex life of the wild boar in Africa, someone in Washington should be able to figure out a way to care for situations like these at a cost that will not pauperize whole families. I'd appreciate hearing from you.

Los Angeles, California

April 15, 1977

NELLIE R. KOSLOWSKI
ALTA LOMA, CALIFORNIA

MARCH 27, 1977

I am not a person who is considered an "at risk" person, but have been closely associated with this disease. My grandfather had this disease, as did two of my aunts and one uncle. Numerous cousins have also been afflicted. I have not had to obtain medical or financial help for such a person but have heard what a difficult time it is to do so.

I really feel it is important to continue the work this Commission does so it will help families which are closely affected.

April 15, 1977

BETTY M. KRONENBERG
LOMPOC, CALIFORNIA

FEBRUARY 27, 1977

I am writing this letter in the hope that it may, in some small way, help the victims of H.D. and their families get the help that they most desperately need. We are an H.D. family. My husband is the victim, and my two daughters, ages 13 and 9-1/2, being at-risk.

When I first met my husband in 1962 he was just a nice, average man, inclined to be rather quiet and easy-going. He was serving in the United States Air Force in England, where we met. We eventually married and had our first child. Ed was having to work a second job at the Base N.C.O. Club (where I also worked part-time) as he had been married previously, and was supporting two older children. Several times when we both worked together, I noticed that while standing he seemed to be swaying and would almost lose his balance. He also had terrible migraine type headaches. I attributed these things to fatigue and excesses of alcohol.

As time passed by, I realized that the happy-go-lucky attitude that I had so much admired was more and more looking like irresponsibility and that we were getting deeper and deeper into debt. At one time we were spending more on alcohol than on groceries.

Within four years we had added two more to our family. Another little girl of our own, plus Ed's older daughter. Ed's nervousness began to show itself in temper outbursts and inability to get along with workmates. Also his headaches had become more and more severe, and he frequently spent days in bed sleeping them off. These days, the children and I would spend creeping around the house, afraid of waking him and invoking his wrath. He was also so nervous at times that he was unable to do some things, such as knocking in a nail, or picking up small objects.

After serving 20 years in the Air Force, my husband retired in September of 1969 at the age of 37. He then went to work on a job training program as a mechanic, and had a dispute with the owner of the business. After this he became a custodian with the Lompoc School District.

One evening after work, he came home with what we thought were the symptoms of a heart attack, which turned out to be pneumonia. I drove him to the Base Hospital Emergency Room, where he was seen by a Dr. Barbarosh. The doctor, after examining Ed, drew me aside and asked me what was wrong with my husband. I really didn't understand his meaning, until he clarified himself by asking me if Huntington's disease was in the family. He then suggested that Ed should see a neurologist.

A few days later, my father-in-law, who had been married to Ed's mother during the early part of her illness, told me that he feared Ed had H.D. The name, Huntington's disease, did have meaning for me, as Ed had mentioned that this was the illness his mother had died of. He had been told by members of a hospital board in Dayton, Ohio, where his mother was, that the chances of him or his offspring getting H.D. were one in a thousand! This was in 1961. I think this illustrates the need for more genetic counseling. After this time, my husband fathered two more children, and already has three grandchildren. The number of people that we can trace as being at-risk from Ed's mother alone is 20. We haven't been able to trace his grandmother's or great-grandmother's families. So just in our family the numbers are enormous and growing.

Well, we eventually did see a neurologist, a Dr. Kenton at March AFB, a 200-mile drive from here, and went back a second time for an EEG. We were to go back again for a psychiatric evaluation, but Ed's condition was getting too severe. Driving that far with him behind the wheel was becoming a nightmare. Anyway, the doctor had told us that he couldn't be sure at this stage, so we were still no wiser. These records incidentally we needed later for a VA claim, but have been lost by the Air Force. Here again, the need for an early diagnosis is apparent.

Meanwhile, the atmosphere at home was getting worse and worse. Ed's temper didn't improve, and he was drinking more and more. The only time we had peace was while he was at work. Like many other spouses of alcoholics, I felt that the problem was just his drinking, so I continually begged him to give it up, or to at least cut down. Of course, as long as he knew I would stay, it did no good. I finally decided that in order to leave, I would need a job. So I went back to high school to get my GED and some job qualifications. A year later, I had my GED, but I also realized that the problem was more than

alcohol. I talked to my in-laws who gave me the name of a man to call in Goleta who was with CCHD. This man proved to be of great help to me, as talking to him made me realize that we were not alone in our problem. It also helped me gather courage to tell Ed what I feared was wrong with him. It isn't easy to have to tell a person that it's possible they have H.D. Strangely enough, the day I picked to tell him was the day a policeman had tried to pick him up on a drunken driving charge.

Well, in September of 1974 we eventually went down to the Wadsworth VA where all it took was for my husband to walk into the doctor's office. His condition was so obvious by this time! Shortly after he spent two weeks at Wadsworth for further tests, after which he came home in great humor for a change. Unfortunately this good state of affairs was not to last, and the drinking soon got underway again. Also at this time Ed stopped work, although he was for a while able to go to school under the GI Bill. The latter, fortunately, coupled with his Air Force retirement pension, allowed us to keep up house payments and existing bills, and live, rather frugally, until the six-month waiting period for eligibility for Social Security was up. How people manage who do not have these benefits to fall back on, I can't begin to imagine!

At about the same time that we applied for Social Security, Ed applied for Total Disability to the Veterans Administration. Well, so far we haven't had any luck with this. Although H.D. cannot really be classified as a service-connected disability, other illnesses of this nature, such as MS, are termed as such and can be claimed as service connected if onset is discovered within five years of honorable discharge. We have since collected numerous depositions from people who knew Ed during his latter service years, to support his disability claim. We are still waiting for the claim to be approved, almost three years later. The fact that all records of us going to March AFB in an attempt to get diagnosis were lost, has been a great hindrance to us. I personally feel that the same five-year rule should apply to H.D. victims.

Meanwhile, for the time being, we are managing financially with our Social Security and the Air Force retirement. Fortunately our house payments are low (living in an area with higher expenses would be more difficult). We are able to go to Vandenberg AFB for the majority of our medical care. As yet we are able to visit a neurologist in Santa Maria at our own expense.

Of course, life in our household is far from easy. I find that a teenager, a pre-teen and an H.D. parent just don't mix. We live a life of constant compromise, and of trying to do anything for a quiet life! There is not a great deal of laughter in our house. Sometimes we spend a whole day of peace. This is something to be very thankful for because the next three days may be filled with anger. Really we could all use a vacation, but how can we? This is an area in which some kind of Government financed respite care would be of great help.

Fortunately, Ed has now stopped drinking. In the end I had to join Al-Anon to help me cope with the problem, and about 10 years too late he finally stopped. I do feel though that in our case the alcoholism occurred during Ed's early service years, in fact, long before the onset of H.D.

Now what of the future? Well, it seems that eventually hospitalization will be the only answer. But the question is, where? Our income is just too high for MediCal to be of much help, even if a hospital close to Lompoc were available. The Air Force can offer no long-term help. The other alternative is the VA Hospital in Los Angeles. The latter is really too far for visiting more than once every week or two. I am afraid I am just not a good freeway driver. To move closer to the hospital is another solution, but would be economically disastrous. Besides that, both of our children have severe allergies, and the older girl has asthma. So a move to a city of smog, and away from our free weekly allergy shots, and Vandenberg AFB Hospital is impractical.

Although we do go to a psychiatrist at our local mental health office once a month to talk over our emotional and family problems, these talks have no real lasting effect. Our oldest child is already experiencing emotional problems from the pressures at home, and I frequently have to talk with teachers and counselors, and explain to them the problems she is encountering at home. Just how long will it be before her problems are more serious than a little natural rebellion at school? She has to let off steam somewhere and home certainly isn't the place. In fact, at home, she spends 75% of her life hiding in her room.

What of the patient himself? In my husband's case he has, from an able bodied breadwinner, able to turn his hand to almost any task around the house, a great mechanic and a good husband and father, become a semi-invalid at 45 years of

age. He still manages to bathe, feed and dress himself, with difficulty. He can barely sign his name, and finds it difficult to read. His days are spent either sleeping or watching television. What kind of an existence is this?

So you can see that there are many ways that us H.D. families need help. This is just part of the story of our family and its problems, so far. I know we probably have much more ahead. Let's hope we have the strength to cope with it all.

Los Angeles, California

April 15, 1977

KENNETH M. KRUEGER
TORRANCE, CALIFORNIA

MARCH 27, 1977

Meeting the family that I am about to tell you about was in 1938 and was a family of six. Father, mother and four children, two boys and two girls. At that time there was no sign of H.D. In 1947, I married the youngest of the girls. At that time my wife's mother was showing definite signs of H.D. However, she was misdiagnosed and was placed into a mental institution. Having lived in the institution for some 15 years, she passed away. After our request, the doctors performed a brain autopsy, proving she has H.D. At this time the oldest child, a girl, was showing signs of H.D., later the youngest boy was stricken, and last was my wife. Only one out of four children has no signs of H.D. He is now 51 years of age.

After my wife watched her sister developing a pattern after her mother's, she became very concerned; however, nothing was mentioned until the passing of their mother. Later, when my wife expected she may have H.D. but would not have an examination, but decided to go back to work to prove to herself she was perfectly healthy. Later I found she had gone through an awful lot of pain and embarrassment due to falling and dropping things all the time, also being asked if she was ill. She finally quit work and later had an examination. The doctor confirmed the fact that she did have H.D. She at that time applied for Social Security, only to be turned down. The reason, she had less than a quarter to go to make it legal for Social Security help. If she hadn't worked that last time she would have qualified. I certainly would think the Government would have different provisions in cases like this.

Being in a large family having H.D., and nine children as possibles, hope the Government will help in research for the answers we're anxious to hear. Along with research, I feel we need homes for the cases where people with H.D. can no longer help themselves. Only we who are involved can vouch that we cannot possibly afford to have our people in private convalescent homes. Another thought about having our own homes, the people would be available for research purposes, for now our doctors and nurses are traveling from one end of the state to the other. One doctor called us one Sunday morning; she wanted

blood samples. She apologized, being Sunday, but mentioned she had a very tight schedule to meet. This was fine with us. When she arrived, she went on to mention she had started in Northern California the first of that week and was going to San Diego from our house, then back home to Colorado.

Thank you for reading my letter and hope it will help in our fight.

I am Kenneth M. Krueger, 54, and married to Ruth L. Krueger, H.D. victim.

Family rundown: Dr. John Lechner, no H.D., father. Edna C. Lechner, H.D., mother. Betty Chambers, oldest child, H.D. John R. Lechner, Jr., no H.D., second child. Donald P. Lechner, H.D., fourth child. Ruth L. Krueger, H.D., third child.

Los Angeles, California

March 15 , 1977

ROBIN LEA KRUEGER
HERMOSA BEACH, CALIFORNIA

MARCH 28, 1977

Since H.D. has afflicted my family, I feel I must make you aware of some of the problems confronting us. My mother was stricken ten years ago. Some of the problems that have arisen since are:

1. Physical limitations, difficulty walking, unable to drive, etc.
2. Social impact on victims and friends, lack of knowledge among society mistaking H.D. patients to being in a drunken stupor.
3. Personality changes, a feeling of failure to communicate.
4. Financial impact, which is the largest problem in my mother's case. She was refused Social Security benefits due to being \$50.00 short the requirement.

There are needs that we have that we alone cannot meet, such as reliable diagnostic tests, obtaining information, guidance, medical assistance, etc., further knowledge to others, and above all, a need for Government-paid rest homes for patients no longer able to help themselves.

Your concern is appreciated. Thank you.

Los Angeles, California

April 15, 1977

MRS. LENA KUEHN
RICHMOND, CALIFORNIA

MARCH 24, 1977

My personal involvement with the ravages of Huntington's disease by having my husband felled by this little-knowncrippler moves me to urge your Commission to do everything you can to seek out a cure. And my third oldest daughter of five daughters was taken about twenty years later, leaving a son and daughter as possible victims. So my plea is doubled.

Los Angeles, California

April 15, 1977

DOMENIC G. LAMPEDECCHIO
PACIFICA, CALIFORNIA

MARCH 5, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

VINCE LAMPEDECCHIO
PACIFICA, CALIFORNIA

MARCH 5, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

April 15, 1977

JUDITH A. LECHNER
FOR: DONALD P. LECHNER
TORRANCE, CALIFORNIA

MARCH 22, 1977

Do you know the agony and heartache of finding out your husband at the age of 37 years old has a horrible hereditary disease called Huntington's disease?

I was 34 years old at the time with three beautiful children and I knew I must tell them of the disease. How do you tell them?

I cannot count the nights I cried myself to sleep worrying about how to tell my two young teenage daughters and my young son. I had to tell them. They accepted the information very well--we cried a lot. But afterwards, they loved their father so much, they did not worry about themselves, all they did was want to make their Dad happy.

Of course, we have problems. My husband, Don, is now 46 years old and hasn't worked for six years. I've been working for 11 years to help support our family. He also has some disability insurance and a retirement from Public Employees Retirement System.

Unfortunately, at the school where my husband worked he did not carry Social Security benefits. When he has to go into a nursing home, I don't know what I will do, because without Social Security he has no Medicare for help in paying the monthly payments. We plan to keep him home as long as possible, but when hospitalization is necessary, I don't know what we will do.

Another thing, I have to get special car insurance, even though I have never had a ticket or an accident, because my husband has Huntington's disease. They say there is always the chance he may try to drive. He has a difficult time just walking--he'd never try to drive a car again.

One of the most helpful things the Commission could do is give us good and inexpensive nursing homes. But most of all, money for research.

Los Angeles, California

April 15, 1977

Please give my family--three children and one grandchild--
some hope for the future. Let's find something to cure or
help people with Huntington's disease.

MRS. PHYLLIS LEWIS
BERKELEY, CALIFORNIA

FEBRUARY 25, 1977

Last Saturday I attended a memorial service of a dear friend who died from the ravages of Huntington's disease. I have seen her weekly for 20 years and watched the disease take its toll of anguish for her, her husband and their four children. She has required 24-hour care for the past 4-5 years. Her four children, ages 25-31, each have a 50% chance of getting this disease. The onset of her disease began at about age 36. It began with a personality change--a lovely, positive person turned critical and became very hostile. The horrible physical symptoms followed.

These four young people, her children, live with the dread that they too will get Huntington's disease. The father too is heartbroken about it all.

He was able to pay the \$800 a month for her care at home. Very few people would have the money or be able to stand the terrible psychological and physical strain to keep such a sick person at home. They had very great love for her and the needed strengths.

Money for basic research for genetic diseases is needed to be given to the National Institutes of Health. We laymen will try to help the families with the disease but please, National Institutes of Health, provide the research money.

Los Angeles, California

April 15, 1977

BRAD LINDSEY
STRATHMORE, CALIFORNIA

MARCH 22, 1977

My father was a victim of Huntington's disease. I am at risk and therefore feel very strongly that Federal funding is needed.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

CARRIE LINDSEY
STRATHMORE, CALIFORNIA

MARCH 22, 1977

My husband was a victim of Huntington's disease. I am very concerned for my children and feel that Federal funding is desperately needed.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

DEBI LINK
AUGSEURG, WEST GERMANY

DECEMBER 10, 1976

My mother is an H.D. patient and my folks live in Villa Park, California. I am 22 years of age and am studying in Germany.

As far as my attitude towards the possible onset of H.D. in myself and my brother and sister, it is something that I reflect upon infrequently. I've observed in myself that when I dwell on the thought extensively, I begin to view normal mistakes or lack of memory as a sign that "I have H.D. and the symptoms are beginning to appear."

At this time in my life, the thought of not having children due to H.D. does not bother me. I must admit that I question as to whether anyone would want to commit themselves to marriage with an H.D. victim. For this reason, my brother, sister, and I are all pursuing careers that offer us a challenging and interesting future with the possibility of not having a marriage partner.

It is difficult for me to visit with my parents more than a month, for I commence to view my mother as a reflection of how I will possibly be and am therefore weakened, depressed, disillusioned (whatever adjective I use will not describe my feelings).

I pray that some medication will soon be discovered that will prevent the illness from impeding my mother's coordination more than it already has. If there is any cure found, please notify the at-risk victims.

Los Angeles, California

April 15, 1977

MRS. JOHN LITTLE
GARDEN GROVE, CALIFORNIA

FEBRUARY 5, 1977

My son was 14 years old when we were told his father had H.D. I had never heard of the disease before that. That same evening his father took gopher poisoning, but it was too slow and too painful; he was found leaning on the horn of his car with all the doors locked. He was rushed to the hospital where his stomach was pumped out. He had permanent liver damage, but he survived. After that he was arrested for drunk driving (it was the H.D. as he didn't drink), could not hold a job, he pawned everything he owned and when that ran out, he started writing bad checks.

He signed himself to Norwalk State Hospital, but that was no help for him, so he signed himself out, rented a motel room and shot himself in the head. For 12 years now, I have lived in fear that our son will get the disease. The hardest thing to do was to tell him he is at risk. It was like pronouncing his death sentence.

Now we both live in fear. My son is now 26 years old and I have noticed the twitching that his father started with. I cannot bring myself to tell him because I could be wrong, but I don't think I am. I want to spare him as long as I can as there is no early treatment that can help at this point. I sometimes think that he has noticed it also, and doesn't say anything, that he may spare me.

My story is probably nothing compared to some of the facts of other H.D. families and patients. I have seen many H.D. patients as I work with Mary Boyle of CCHD and in doing so, I have decided that unless they find a cure or a way to control this horrible disease, I love my son too much to have him go through life like that. I would hope for him to follow his father even if it means suicide.

Thanks so much to Marjorie Guthrie for calling this disease to the attention of so many people, and hopefully the right people.

I would like to know the place of your meeting to be held in California on April 15 of this year.

Los Angeles, California

April 15, 1977

MRS. E. L. MacINTOSH
CUCAMONGA, CALIFORNIA

MARCH 27, 1977

Having been acquainted with Huntington's disease since early childhood, I feel impelled to request that the study for control and cause of this horrible disease be of the most urgent necessity.

In early 1930s an aunt lived with us in Lincoln, Nebraska and I vividly remember the disease, and a most wonderful doctor who tried in vain to get help in any way for all those concerned. It is only by the grace of God that neither I nor my children need not worry of being a risk in this terrible thing.

Having seen aunts, uncles and cousins suffer the agony and humiliation, again I urge the importance of this Commission. Incidentally, the doctor's name was Dr. Royal.

Los Angeles, California

April 15, 1977

MR. AND MRS. A. E. MAFFLY
BERKELEY, CALIFORNIA

MARCH 1, 1977

We urge your Commission to give all its support and make every effort to cure Huntington's disease. It causes such suffering for the victims and their families and for future generations.

CHARLES H. MARKHAM, M.D.
PROFESSOR OF NEUROLOGY
UCLA SCHOOL OF MEDICINE
LOS ANGELES, CALIFORNIA

JULY 15, 1977

The solution to the problem of Huntington's chorea lies in three interlocking, simultaneous efforts: research, treatment, and education.

There are two main research approaches which may pay off:

1. Identification of the basic genetic enzymatic defect. This will take scientists working in seemingly unrelated fields who actually know little about Huntington's chorea. Finding and involving the "right" scientists is a matter of education and money for grant support. Once the basic enzymatic defect is found, there are three main approaches to treatment:

(a) Replacement therapy of the specific agent or its immediate precursor.

(b) Circumventing the metabolic pathway which is blocked for lack of the specific enzyme. Example: low phenylalanine diet in phenylketonuria, where the problem is a failure to convert phenylalanine to tyrosine because of a lack of a specific enzyme, phenylalanine hydroxylase.

(c) In utero detection in pregnant women of the lowered level of specific enzyme or the piling up of its metabolic substrate. Example: accumulation of G_{M2} gangliosides in Tay-Sachs disease.

2. Clarify the structure and function of key areas involved by the disease. The neostriatum (caudate and putamen) and cerebral cortex are prime areas for such consideration. Particular emphasis needs to be placed on neuron interconnections, and on neurotransmitters and neurohumors, the chemical substances cells use to communicate with each other. Treatment follows directly from this:

(a) Reduce the amount of transmitter which aggravates certain symptoms.

(b) Increase or keep at an effective level for a longer period of time a transmitter which is reduced in amount. Example: DOPA in Parkinson's disease.

Education is crucial for the ultimate solution of Huntington's chorea. There are two main audiences.

1. First are the patients, their families and friends. They must know the hereditary pattern of the disease, what a dominant pattern of transmission means, and that at the present time the only sure prevention of the disease is for "at-risk" individuals not to have children. Education will assume even greater importance when the disease can be detected in utero. Again, keep in mind the example of Tay-Sachs disease.

2. The second group which needs education are the scientists who are working in areas which may have relevance to Huntington's chorea--but they don't know it. A most important vehicle to reach these scientists are the interdisciplinary workshops sponsored by the H.D. Foundation.

In conclusion, a concentrated push in basic research plus education has a good chance of leading to effective treatment or prevention of this devastating disease. These approaches will take not only money but an active and continuing interest on the part of all affected individuals and their elected and appointed Government representatives.

April 15, 1977

GERALD P. MCAULEY
BANNING, CALIFORNIA

MARCH 8, 1977

Relative to the hearing by your body scheduled to be held on April 12, 1977, I have been requested to relate to you my family experience with Huntington's disease and am only too glad to do so in the hope that the results of your hearing may prove to be an opening wedge in the eventual eradication of this most dreadful disease. The University of Michigan conducted an investigation of the disease about thirty years ago and requested all the information we could give them as to our family history. Not knowing just exactly what it is that you wish to know, I will repeat that information in regular type and relate the consequences of it in capital type so that you may more readily disregard the family history if you wish to do so.

My name is Gerald Patrick McAuley, born November 6, 1900, in the city of Watertown, New York. My present address is 1738 N. Alessandro, Banning, California 92220.

Grandparents on my father's side were Charles McAuley, born in County Antrim, Ireland. Dates of birth and death not known by me. His wives were Mary Bannon, nee Bourke, born in Queens County, Ireland, dates of birth and death not known by me; the name of his first wife and the dates of birth and death are not known by me. The first wife of Charles and the husband of Mary were lost in a shipwreck on the way to Canada. Charles and Mary arrived in the eastern part of Canada probably between 1820 and 1840 and were probably married in the vicinity of Spencerville, Ontario.

The children born of Charles and Mary were: John, married to a Margaret Lahey; Walter, married to Mary Ann Kelly; James, married to Rose McLarry (my mother); Charles, a twin brother of James was married late in life but not known to whom; David was single; Mary was married to a Lahey, first name not known; Catherine was married to a Liddy, first name not known; Elizabeth was married to James Nolan; Julie was married to a man named Wolf. Most of the children were probably married in the vicinity of Spencerville, Ontario. It is suspected that David was a victim of infantile paralysis, at least he was crippled. IT IS BELIEVED THAT NO CHILD BORN OF THE ABOVE UNIONS BECAME A VICTIM OF HUNTINGTON'S CHOREA OTHER THAN JAMES AND ROSE McLARRY. (THEMSELVES NO, CHILDREN YES.)

Grandparents on my mother's side were: Patrick McLarry born in Londonderry, Ireland, 1815, died 1898. His wife was Rose Weir, born in County Antrim, Ireland, date not known, died in 1910.

Children born of the above union were: Rose, married to James McAuley (my mother and father); Kate, married to Andrew Buckham and later, after his death, to John Rainey; Jack, married to Nancy Armstrong; and Mary, Margaret and Charles, all three single. IT IS BELIEVED THAT NO CHILD BORN OF THE ABOVE UNIONS BECAME A VICTIM OF HUNTINGTON'S CHOREA OTHER THAN JAMES AND ROSE McLARRY.

James and Rose had ten children: (1) Wilhelmina, born in Kemptville Parish, Ont., August 18, 1879, died Feb. 18, 1969 in Detroit, Michigan; (2) Charles, born in Ventnor, Ont., Oct. 14, 1882, died in Dearborn, Michigan, May 16, 1954; (3) George, born in Groveton, Ont. December 25, 1884, died in Detroit, Michigan, Sept. 25, 1957; (4) Margaret Helena, born in Groveton, Ont., September 22, 1886, died in Windsor, Ont., Sept. 25, 1968; (5) David L., born in Groveton, Ont., Jan. 26, 1889, died in Gulfport, Miss., July 4, 1956; (6) Leon S., born in Groveton, Ont. June 22, 1891, now residing in Banning, Cal.; (7) James F., born in Groveton, Ont. April 13, 1893, died in St. John, New Brunswick, 1972; (8) Cecelia, born Feb. 6, 1881 in Willows, Cal., died June 6, 1950, in Detroit, Mich.; (9) John H., born in Cardinal, Ont., Feb. 26, 1898, died July 4, 1946, in Nankin Township, Mich.; (10) Gerald P., born Nov. 6, 1900, in Watertown, N.Y., now residing in Banning, Cal.

Of the above children only David L., James F., and Gerald P. were married.

ALSO OF THE ABOVE CHILDREN, ONLY JOHN H. AND DAVID L. BECAME VICTIMS OF HUNTINGTON'S CHOREA. THE FIRST SIGN OF THE DISEASE IN JOHN H. WAS TROUBLE IN LIGHTING A CIGARETTE. HE WOULD, AFTER STRIKING A MATCH, BRING IT TO THE END OF THE CIGARETTE WITH A WIDE CIRCULAR MOVEMENT WITH HIS RIGHT HAND. LATER HE BEGAN WALKING LIKE A DRUNKEN PERSON. THIS GOT STEADILY WORSE TO THE EXTENT THAT HE WOULD ONLY GO OUT OF THE HOUSE AFTER DARK, AND THEN NOT FOR VERY LONG. HE THEN BECAME SO SELF-CONSCIOUS THAT HE, WITH THE HELP OF A BROTHER, BUILT A MAKESHIFT ROOM IN THE BASEMENT WHERE HE LIVED AND SLEPT, AND WHERE HIS MEALS WERE BROUGHT TO HIM, REFUSING TO EAT WITH THE REST OF US UPSTAIRS. HIS SPEECH BECAME VERY BLURRED AND UNINTELLIGIBLE. FINALLY, BECAUSE HE COULD NOT CARE FOR HIMSELF,

AND BECAUSE OUR OLDEST SISTER COULD NOT DO SO, HE WAS ADMITTED TO THE COUNTY HOSPITAL IN NANKIN TOWNSHIP WHERE HE UNDERWENT SHOCK TREATMENTS IN ICE COLD WATER, AS A RESULT OF WHICH HE CONTRACTED PNEUMONIA AND DIED.

When David L. was about 28 years of age, he married Eva Donohue of Watertown, New York in Detroit, Michigan. They had seven children: Robert, John, Regina, Mary, Laurita, Joseph and Ellen.

DAVID L. BECAME A VICTIM OF HUNTINGTON'S CHOREA AT BETWEEN 45 AND 50 YEARS OF AGE. HE WOULD HAVE THE SAME TROUBLE LIGHTING A CIGARETTE THAT JOHN H. DID. HIS SPEECH ALSO BECAME BLURRED AND HESITANT. I DID NOT SEE DAVID VERY OFTEN DURING THOSE YEARS, BUT EVENTUALLY HIS WIFE DIVORCED HIM. IN 1945 HE WENT TO GULFPORT, MISS. TO SEE HIS SON, JOHN, WHO WAS IN THE NAVY. IT IS THOUGHT THAT WHILE THERE, HE BECAME A VICTIM OF A ROBBERY AND MURDER BY DROWNING AT THE HANDS OF PERSON OR PERSONS UNKNOWN.

OF THE CHILDREN BORN OF DAVID L. AND EVA, ROBERT, JOHN, JOSEPH, AND ELLEN BECAME VICTIMS OF HUNTINGTON'S CHOREA.

From this point on, I do not have any dates of events occurring. After being discharged from the Army of World War II, Robert married a girl by the name of Leona, last name not known to me. They had one child by the name of Donna Mae who is now married but to whom I do not know.

I DON'T THINK ROBERT WAS MUCH OVER FORTY YEARS OLD WHEN AFTER AN AGONY OF FRUSTRATION ON THE PART OF HIS WIFE, HE WAS ADMITTED TO THE VETERANS HOSPITAL IN BATTLE CREEK, MICHIGAN. ON OUR TRIPS BACK TO DETROIT FROM CALIFORNIA, WE WOULD MAKE AT LEAST ONE TRIP TO BATTLE CREEK TO SEE ROBERT. AT FIRST HE WAS PERMITTED TO TAKE A WALK ON THE GROUNDS WITH US, DURING WHICH TIMES HE WOULD TELL US THAT HE HAD SEEN MY BROTHER, JOHN H., AND HIS FATHER FLOATING AROUND IN THE AIR OVER THE GROUNDS. HE WOULD TELL US THAT HIS WIFE HAD COME TO VISIT HIM IN A BRAND NEW CADILLAC AUTOMOBILE AND BE SO PROUD IN TELLING IT. LATER, HE WAS NOT ABLE TO RECOGNIZE US AND HIS SPEECH WAS SUCH THAT WE JUST COULD NOT UNDERSTAND WHAT HE WAS TRYING TO TELL US. AT THIS STAGE OF HIS ILLNESS HE WAS NOT PERMITTED TO GO WITH US FOR A WALK ON THE GROUNDS. HE ALSO USED TO TELL US THAT HE WAS THE ANZIO KID, REFERRING TO THE BEACHHEAD LANDING AT ANZIO; IN FACT, HE WAS KNOWN AMONG THE OTHER PATIENTS THERE AS THE ANZIO KID. POOR BOB, SUCH A GREAT GUY, AND SUCH A GREAT PITY

TO HAVE TO DIE AS THE RESULT OF A DISEASE WHICH OUR VERY BEST DOCTORS ARE NOT ABLE TO PREVENT OR TO CURE.

ANOTHER TRAGIC CASE IS THAT OF HIS BROTHER, JOHN, MOSTLY REFERRED TO AS JACK. AS A TEENAGER HE WAS AN AMATEUR BOXER AND WAS PRETTY GOOD AT IT. WHEN HE JOINED THE NAVY HE WOULD ENTERTAIN THE CREW BY FIGHTING THE CHAMPIONS OF OTHER SHIPS AND FOR HIS WEIGHT CLASS, BECAME CHAMPION OF THE PACIFIC FLEET. UPON HIS RETIREMENT AS CHIEF PETTY OFFICER, HE WAS EMPLOYED BY A TITLE SEARCH COMPANY IN SAN DIEGO. HE WAS ALSO A PRETTY GOOD CHESS PLAYER, TAKING PART IN TOURNAMENTS AND WINNING A FEW. HE WAS IN GOOD SHAPE, BOTH PHYSICALLY AND MENTALLY. AFTER A FEW YEARS WITH THE TITLE COMPANY, THE DREAD HUNTINGTON'S DISEASE BEGAN TO MAKE ITS INROADS UPON HIM. HE ASKED HIS BOSS ONE DAY IF THEY WANTED HIM TO RETIRE. THEY THEN ASKED HIM TO HANG IN THERE UNTIL HE COULD NO LONGER DO HIS WORK. IT GOT SO HE COULD NO LONGER DRIVE HIS CAR AND AS A CONSEQUENCE WAS FORCED TO AGAIN RETIRE. BECAUSE OF A LACK OF CONCENTRATION OR LOSS OF IT, HE WAS ALSO FORCED TO GIVE UP CHESS AS A HOBBY. HE DOESN'T TALK MUCH ANY MORE. WHEN HE TRIES TO WALK, HE RUNS AND RUNS INTO THINGS IN THE HOUSE, KNOCKING OVER LAMPS AND CHAIRS OR ANYTHING THAT GETS IN HIS WAY, EVEN HIS WIFE. HE FALLS A LOT WHICH REQUIRES THAT HE BE HELPED TO HIS FEET AGAIN WHICH IS BEGINNING TO BE AN ALMOST IMPOSSIBLE TASK FOR HIS WIFE, POOR GIRL. SHE TELLS ME THAT SHE IS GOING TO WRITE TO YOUR COMMISSION ABOUT HER EXPERIENCES SO I WON'T BOTHER SAYING ANYTHING MORE ABOUT HIM. SHE CAN TELL YOU BETTER THAN I CAN.

About Joseph, who lives in Florida, and Ellen, who lives in Detroit, both of whom are married. I am not able to furnish any information other than to say that I hear through the grape vine that they are both in pretty bad shape, and that they both have children.

Because Huntington's chorea is supposed to be an hereditary disease, the University of Michigan and other persons with whom I have talked, cannot seem to understand how my mother and father were able to escape it, and, supposedly, pass it on to two of their children. I only know that neither of my parents showed any signs or indication of the disease.

TO GET BACK TO JACK AND HIS WIFE, DOROTHY, FOR JUST A MOMENT, SHE IS HAVING ONE HELL OF A TIME TRYING TO GET HIM ADMITTED INTO A VETERANS HOSPITAL FOR THE KIND OF CARE AND PROTECTION WHICH SHE, PHYSICALLY, IS NO LONGER ABLE TO PROVIDE.

Los Angeles, California

April 15, 1977

SHE HAS ALREADY SUSTAINED A BROKEN RIB AS A RESULT OF TRYING. IT CONCERNS ME GREATLY THAT SHE MAY AGAIN BE INJURED, AND THAT HE MAY ALSO INJURE HIMSELF EITHER ACCIDENTALLY OR ON PURPOSE TO THE POINT OF DEATH. HE IS SO PROUD AND FEELS SO HOPELESS. PLEASE, PLEASE, HELP THEM; AND ALL OTHERS WHO HAVE THE DISEASE.

It is my sincere hope that what I have related to you will be of some help in the deliberations of your committee.

Los Angeles, California

April 15, 1977

MR. AND MRS. FRED C. McCREAKEN
REDONDO BEACH, CALIFORNIA

MARCH 17, 1977

I am writing in regard to an acquaintance of mine, who has Huntington's disease. Her name is Ruth Krueger. I feel that something should be done on a nationwide basis. Perhaps the Government could help financially. A rest home would be most beneficial as their condition tends to become worse with the passing of time.

She has tried to get financial help through the Social Security Disability Program but has been refused.

Hope you can do something for her and the hundreds of more like her.

Los Angeles, California

April 15, 1977

PENNY MENDES
OAKDALE, CALIFORNIA

FEBRUARY 22, 1977

I'm writing this letter hoping it will help urge your Commission to make every effort to combat and cure Huntington's disease.

It is in our family as my father-in-law, who is 56 years old, has been afflicted. We did not know it was even in the family until he was 54 years old and the H.D. was diagnosed. Dad's mother was an H.D. victim and her mother was also, but she died 20 years ago. At that time the medicine and science wasn't as it is today. So now Dad's four children all have children. Thus I am very concerned about H.D. Dad is handling the disease fairly well, but we have our share of problems. Soon it'll be time to obtain professional care in his everyday living and we feel the need for an H.D. facility.

As of now, Dad has become a very stubborn man and we have to deal with him as a child in many cases. His mental attitude is pretty good considering all that he knows that he has to go through.

It is a very destructive and crippling disease. We desperately need an H.D. facility in our area and other areas of this U.S. In the past 2-1/2 years, the research has been very encouraging, but we NEED MORE. It is so hard to sit and watch him suffering but what can we do besides making him as comfortable as possible?

WE NEED MORE RESEARCH AND TRAINING in combating and curing this horrible disease.

I am very concerned too because my husband is an at-risk person along with his sisters and brother. He still has about 15 years before he will know if his body is a carrier, but hopefully by then there will be a CURE!

PLEASE HELP US AND OUR CHILDREN and the many other families afflicted by this disease.

Thank you.

DORIS MERRIMAN
GRIDLEY, CALIFORNIA

FEBRUARY 7, 1977

My only daughter, Jana, inherited a living death from her father. She was born with a time bomb concealed in her brain, set to go off in the very prime of her life.

In her mid twenties, Jana began to show signs of mental illness. Her marriage fell apart and Jana was no longer able to cope. We put her in a mental health program, hoping that therapy would help her back to normal. Instead, there was a steady decline in her condition. She developed a twitching in her face and a peculiar twist in her walk. She became careless about her appearance and slovenly in her dress. Still, the doctor insisted she was schizophrenic and ignored a letter from a hospital which suggested that there was something organically wrong with Jana.

Jana was shifted from one home care unit to another, but as her condition worsened, in desperation we took her to a state institution. There her medication was stabilized and she became calm and rational. Soon, however, she became very disorganized and confused. One of the doctors on the staff became interested in Jana and seemed determined to help her. She decided the problem was organic and took Jana to University Hospital in San Francisco for tests. All were negative and the reports came back diagnosed as schizophrenia with brain damage in the frontal area. Jana returned to the institution.

I cannot describe the sick, empty feeling which came over me--I believe I was actually hoping there was a brain tumor, which I felt we could combat. I think that was the day we lost all hope. I asked Dr. Strathman to send for the medical records of Jana's father who had died in Veterans Hospital in Little Rock, Arkansas. When she received them, she asked me to come for a conference. I had never heard of the disease called Huntington's disease. This devastating illness has been hidden, wrongly diagnosed and denied ever since the first paper was written on it by a Dr. Huntington back in 1872. It is so rare, even Dr. Strathman said she had to look it up in the library to be able to tell me anything about it. By the time our conference had ended, something inside me had died. I have not had a completely happy day in my life since that day and I do not believe I ever will again.

It was months before I found the courage to tell my two grandchildren that they have a 50-50 chance of inheriting this horrible disease. How does one tell young, vibrant people that they must never have children of their own? I believe everything possible must be done to stamp out this disease.

I keep in touch with the National Organization and one year I attended a meeting of the California Chapter in Los Angeles. While it was a great help to be with others who share the same problem, it was also heartbreaking to hear the troubles and indignities they suffer. When recent HEW bills have been vetoed by past presidents, it is indeed a blow to research for neurological diseases. But then, perhaps, after nearly 100 years of nothing being done for the 100,000 afflicted persons of Huntington's disease, it is too much to hope for a scientific breakthrough in the near future. To quote the father of one of the afflicted persons, "When I heard that this nation left 15 million dollars worth of equipment on the moon, I was indignant--damned indignant--15 million would probably solve our problem." The people of this country have always rallied to the support of Polio Drives, Heart Funds and numerous other needs. Why can't we bring this disease out in the open and make the public aware of its devastation?

Jana's normal life was much too short. How sad to see her children deprived of their mother. How sad that such a beautiful young woman is lost to her family and to the world. Jana has suffered so much. She has been overdosed with drugs, denied drugs after she was addicted, locked in rooms, unattended for hours, abused by nurses and staff. Her teeth decayed--no dentist wanted her as a patient. She now weighs approximately 50 pounds, a skeleton still pursued by those uncontrollable movements known as Huntington's disease.

I send you this information in this personal form, hoping to relate to you, some of the heartbreak and despair suffered by H.D. patients and their families.

Thank you.

April 15, 1977

MARY MILLER
TURLOCK, CALIFORNIA

FEBRUARY 23, 1977

Although H.D. is not in my family, I have been close friends with an H.D. family for over ten years. All that I can say is that an appalling situation exists for an H.D. family. In this particular family, the mother of four girls (currently aged 11-24) was the affected member. For years she was diagnosed as mentally ill, in and out of institutions, given no treatment but tranquilizers. She was a financial and emotional burden to her family. No one knew what to do with her. She claimed to have H.D. in her family, but other family members denied it, and it was assumed she suffered from delusions.

For the next 3-4 years the doctors alternately assured the family that she did and didn't have H.D. They ran and reran tests. The family was alternately reassured and alarmed. It was such an ordeal.

Then when it was finally definitely determined she did have H.D., the family became involved in the local CCHD. They became even more discouraged with the hopelessness of the situation. What they found was dozens of families like themselves: no treatment, no cure or hope for one, no money for research. All they could do was hold each others' hands.

Everywhere they turned, doctors, social workers, genetic counselors, all confessed that they knew very little about H.D.

The need for research and family counseling with H.D. victims and their families is crucial. I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

NOAH R. MILLER
TURLOCK, CALIFORNIA

FEBRUARY 17, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

MARY P. MILLS
PALO ALTO, CALIFORNIA

MARCH 26, 1977

How can anyone possibly put into words the agony, despair, frustration of those of us who have Huntington's disease in the family? In the last two days I have typed dozens of pages telling our story, pouring out the anguish, the pain, the end of our dreams and hopes; the despair that brought one of us to the brink of suicide; a mental breakdown; the hundreds of sleepless nights; the waking in the night violently sobbing so that we may keep smiling during the day; the struggle for acceptance and finally feeling that one has "accepted" only to feel the anger again when I see how rapidly my daughter is fading away; the anger at my own countrymen, because my daughter, with her stumbling walk and jerking movements, is accepted completely overseas, but is so often stared at with repulsion here in her own country because she is "different," so she is happier overseas; the feelings of helplessness because there is nothing that can be done. I have learned to accept many things, but the one thing I can never accept is that nothing can be done--if we could only get sufficient funds to go full steam ahead on research--if we could find a treatment.

I cannot send my dozens of pages. You do not have the time to read them. Instead, this: Sometimes I think Huntington's families must be the quietest families in the world. I have attended so many CCHD meetings and they are the quietest meetings I ever attend. People tell their stories quietly, simply, showing great courage, but the room vibrates with their quiet despair. I used to drive home thinking, "CCHD families remind me of something, but what?" Suddenly one night there flashed through my mind the pictures of the concentration camps in World War II--condemned people sitting there quietly, all hope gone--and I had my answer. CCHD families sit quietly with the same expression on their faces, a condemned minority without hope.

And why do we have no hope? Because 105 years after H.D. was first "discovered" there still is no treatment, no cure, still very little research being done.

Why is so little research being done? Because there are no funds for research. We are the forgotten minority. Yes, we are one of the minorities; we do not have a "popular" disease.

I am fully aware that billions of dollars are needed for research for many thousands of diseases, but H.D. has truly been the forgotten, the ignored disease. Are we of so little worth, of so little value in a country that prides itself on the worth and value of the life of each human being?

In my frustrated moments I think we should go marching through the streets with placards, demonstrating, shouting, crying out our agony and pain as so many other minorities have had to do until they finally were heard. Our 100,000--and growing--families, are we of so little worth to you that you can just sweep us under the rug? Because that is what has been happening to H.D. CCHD and interested parties, after years of effort, finally were able to get a Commission appointed. YOU are the Commission and you are now beginning to listen to us. I sincerely hope you are not listening to us. I hope you are hearing us.

Seven years ago I had never heard of Huntington's. A phone call from a distant relative of my ex-husband's (who had disappeared 15 years before) started my world crashing around me. The relative casually mentioned that my ex-husband (and the father of my daughter) had some disease that caused his arms and legs to jerk violently; that he was in some VA hospital; that it seemed to be the same thing his father had had. That is all he knew. I came on full alert that this was something hereditary and my daughter might be affected. My search began. I will not go into the details of that search, but it took me one year from the time of that phone call until the facts and records were in my possession. The verdict: my daughter's father had Huntington's chorea; her grandfather's death certificate read "Huntington's chorea."

What was Huntington's chorea? I had never heard of it. I went to three doctors before I could find one who could tell me anything. My family doctor said, "Oh, it is just a form of St. Vitus' Dance." The second doctor did not know. The well-known neurologist finally gave me all the facts known, clearly, completely, pacing the floor as he recited them. Every word he said is still burned into my soul. He ended with, "I wish I could at least offer you hope, but I cannot even do that. Little research has been done on this disease because funds are not available for study of an unpopular disease and Huntington's is an unpopular disease." He said this with sadness and frustration.

I sat there stunned. I had no idea what a horrible disease we were facing. But I still had hope; my daughter still had a 50-50 chance.

I still remember nothing of the next few hours until I found myself in my living room saying the word "Gethsemane" over and over. Yes, I had been there too.

How could I tell my daughter, who was living in Turkey at that time and excited over starting a family? How do you tell your only child, a bright, sensitive, warm, loving, responsive human being, "Wait! When you were born your parents gave you an unexpected gift, not just the gift of life but a time bomb with a short fuse which may be ticking away inside you." Inside I was screaming, "No, I will not tell her. Since there is no way to make an early diagnosis, since nothing can be done if she does get it, since there is nothing to do but wait, why should I make her life a hell for the next ten years? The doctors tell me she should have at least ten more years before symptoms will appear and that is their only way of diagnosing it." But I could not afford that luxury because they were planning a family and they must have the facts so they could make the choice whether or not to have children.

She did not have the ten years. Less than two years later, just after her 31st birthday, she was diagnosed as having H.D. She has never complained. Her courage and determination to live each day joyously and fully have been an inspiration to all who know her.

She had been a teacher and loved teaching more than anything she had ever done. She is still able to walk, although with great difficulty; her speech is slurred; she is gaunt; she looks very old at times. But I shall be eternally grateful to South Korea for allowing her to teach there in 1975-76 even though she was so weak at the end of the school day that she had to be put to bed immediately. In this, her own country, they would no longer allow her to teach in her condition. The doctors still do not understand how she did it. I do. It gave her a purpose which kept her fighting spirit; it made her feel she was still making some contribution; it proved to her that she could still function, no matter what the cost. The doctors think it speeded up the course of her disease, shortened her life. What life?

From personal experience I know how desperately money is needed for research. Several years ago, although I had a full-time job, I spent many hours of volunteer time working at Stanford on a project being done on Huntington's out of Dr. Linus Pauling's lab. Why? Because the biochemist doing the research poured out his bitter despair to me that he did not have the funds to complete his project and asked if I could help even though I had no background in science. He said, "I feel we are so close to finding something significant. How can we ever find the answers without the funds to help us?" Many CCHD members pitched in to help. But we were not qualified personnel.

I also have done some volunteer work at the VA hospital in Palo Alto under a brilliant and dedicated doctor who is working desperately to find some treatment for his H.D. patients. He, too, desperately needed volunteers to help. Dr. Kenneth Davis is an expert on H.D. I will read a rather encouraging article in some medical journal about some new drug for H.D., ask Dr. Davis, and his answer usually, is "Oh, no, we have already tried that and it does not help." He is still working and still needing money.

Research has begun to come up with some facts, just the beginning stages really, but with more funds--and I mean millions, not thousands--there can be at least some treatment, if not a cure. I still cannot accept the fact that we have no money to work with when I think there is a strong possibility that something can be found for treatment. Something! Cancer patients have helpful drugs, therapy, often cure, and always hope, for so much research is being done on cancer. Huntington's patients have nothing. It is up to you to decide whether we are worth anything as human beings so that you can provide some money to make us feel that our children, our families are worth saving, are worth helping.

Can you imagine how I felt, after years of doing fund drives in this area, when I did my last cancer drive two years ago? People gave generously, as always, but as I left each door, I felt an increasing sadness because I knew I could not say, "Would you please contribute a little for H.D.? We have almost nothing for research. There are millions being collected for cancer, as well as all the government funds available, but there is nothing for the disease from which my daughter is dying." I still do drives for other causes, but can you not understand my sadness?

Knowing there comes a time when H.D. patients can no longer be cared for at home no matter how much you want to keep them, I have also researched the Bay area and the Peninsula area to find hospitals, extended care facilities, etc. that would take H.D. patients. I found not one except Napa Hospital, a state mental institution. Some H.D. patients become demented and may need a mental institution, but many minds are clear and they do not belong in a mental institution. Let's face it. THERE ARE NO FACILITIES FOR H.D. PATIENTS. We do not want to put them into hospitals and homes, but when the time comes when they can no longer be cared for at home, where do they go? Please tell me. Where do they go?

I live on Social Security and a small pension and I intend to keep my daughter at home for so long as I can stand on my two feet, but even if I had millions of dollars, could I find enough nurses to care for an H.D. patient at home? I have talked to nurses too; there are not many who want to care for an H.D. patient.

I am but one voice. You have the power. Are you hearing us? Can you not help those of us who are despairing and desperate? Can you not at least give us a chance for hope that money will be forthcoming? I am asking only for a fighting chance and we need money for that fighting chance.

Los Angeles, California

April 15, 1977

MRS. VICTORIA MOORE
TORRANCE, CALIFORNIA

MARCH 23, 1977

I am writing in regards to my neighbor who has H.D. Her name is Ruth Krueger. I've seen her trip and fall many times. I know the whole family and feel like something should be done; perhaps the Government could help financially. They could really use a rest home as their condition gets worse as time goes on.

I know they are not mentally ill and do not need a mental institution, could not afford any kind of a rest home.

I have read all her correspondence with Social Security disability to no avail.

Hope you can do something for her and hundreds of more like her.

Los Angeles, California

April 15, 1977

MRS. K. MURDOCK
MOUNTAIN VIEW, CALIFORNIA

MARCH 10, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

PEGGY MUZNY
FRESNO, CALIFORNIA

MARCH 22, 1977

I am friends with a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

BETTY NAKAYAMA
FRESNO, CALIFORNIA

MARCH 22, 1977

I am a friend of a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

ERMA NATION
MODESTO, CALIFORNIA

FEBRUARY 14, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

JERRY L. NATION
MODESTO, CALIFORNIA

FEBRUARY 14, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

KENNETH W. NATION
MODESTO, CALIFORNIA

FEBRUARY 14, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future families, I urge your Commission to make every effort to combat and cure it.

April 15, 1977

JANET Y. NULL
GARDEN GROVE, CALIFORNIA

FEBRUARY 25, 1977

Several years ago my brother became ill. He was unable to think straight, to remember things, and it became difficult for him to speak and walk. He also had a "family trait" (like our father and grandmother); his body would twitch and jerk. Six years passed before his illness was properly diagnosed as Huntington's disease. (By this time he had lost his job, his wife had to go to work, etc.)

When the diagnosis was made, the first thing we tried to do was find out, "What is Huntington's disease?" We discovered that the information on H.D. was quite limited and frightening. We did find, however:

1. There is no cure.
 2. There is no effective control.
 3. There is no diagnostic test to detect it prior to the onset of symptoms.
- AND:
4. Our father also has H.D. as did his mother.
 5. My sister and I are both "at-risk" and "if" we have H.D. (no symptoms as yet), our children are "at-risk."
 6. My brother's children are "at-risk."

It was extremely traumatic for my family to learn all this and to be faced with such a dreaded disease.

As the disease progresses in our family members, more and more problems continue to arise. Without going into great detail, let me give you an example: Since my brother is at the point where he can no longer live at home, he has been placed in a rest home. Medicare insists that the treatment he is receiving is "custodial care" thus not covered under Medicare benefits. And if Medicare does not cover the treatment, neither will private insurance. My sister-in-law is faced with giving up all their assets (home, etc.) and going on welfare; or divorcing my brother, so she and the children can keep what she and my brother worked so hard for, and putting my brother

on welfare. This, of course, is not a fair choice, but where can she go and what can she do to alter this situation?

From my family's past experiences with Huntington's disease, I feel that:

Qualified professionals are needed to assist H.D. patients and families with legal and financial problems.

Qualified Social Service personnel are needed who are well informed on H.D. as well as the problems connected with it; i.e., insurance, employment, disability, health care, etc.

Qualified psychological services need to be set up to assist H.D. patients and families with the psycho-social problems associated with the impact of H.D., i.e., marriage, family planning, employment, etc.

H.D. information needs to be made available to the medical profession in general so H.D. may more readily be recognized and properly diagnosed. Doctors need to know where specific treatment and research are performed, etc.

H.D. information needs to be made available to health care facilities so they may care for the H.D. patient more effectively.

And, of course, H.D. research should be continued and increased until there are effective controls and eventually a cure.

It is obvious that the needs of Huntington's disease patients and their families can not be met without substantial funds and proper leadership. Hopefully through the H.D. Commission the necessary funds and leadership will be provided by our Government.

Los Angeles, California

April 15, 1977

WILLIAM H. NULL
GARDEN GROVE, CALIFORNIA

FEBRUARY 25, 1977

My wife and I have been working with Huntington's disease families and others interested in helping them. Having H.D. in my wife's family makes us keenly aware of the problems faced by H.D. patients, their families and those trying so desperately to understand and help.

Take this family for example: The mother has H.D. (as did her mother), the father is deceased and there are eleven children, some married with children of their own. Since there is no "breadwinner" the mother is on welfare and those children living close by are doing all they can to help. Their knowledge of H.D. prior to our association with them was that their grandmother died of H.D. and now their mother has it. As they learn more about the disease, they are able to help their mother more effectively, and yet, the knowledge that they, too, are "at-risk" creates even greater emotional problems. The family not only needs physical and financial assistance, but psychological as well.

There are other cases where finances are not a major burden but because of the fear of not being covered by life and medical insurance on their job or even losing their job because they are "at-risk" is a continual burden.

I could continue on and on and the fact would still remain that the H.D. victim, his family and friends need help. I sincerely hope that the Commission will succeed in making our Government aware of this vast need of an ever increasing number of its citizens.

Los Angeles, California

April 15, 1977

BARBARA PAGET
TORRANCE, CALIFORNIA

MARCH 17, 1977

I am writing in regards to an acquaintance of mine who has Huntington's disease. Her name is Ruth Krueger. I feel that something should be done on a nationwide basis. Perhaps the Government could help financially. A rest home would be most beneficial, as their condition tends to become worse with the passing of time.

She has tried to get financial help through the Social Security Disability program but was refused.

Hope you can do something for her and the hundreds of more like her.

Los Angeles, California

April 15, 1977

MARJORIE PANNELL
RIPON, CALIFORNIA

MARCH 7, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

ALMA E. PARKS

OAKLAND, CALIFORNIA

MARCH 7, 1977

Our daughter died of Huntington's disease brought on by terminal cancer on January 18, 1975. Although she did not live in Oakland we were able to visit her frequently.

She was just 28 years old and left three children: Kenneth, age 10, Sherrie, age 8, and Timothy, age 3 (which I have recorded in the Northern California Chapter, Committee to Combat Huntington's Disease, 411 30th Street, Suite 601A, Oakland, California 94609).

I am a member and realize that these three precious grandchildren may some day be afflicted with this dread disease for which there is no cure at this time.

Our daughter (adopted) suffered untold agonies and pain not only in the advanced stage of Huntington's disease but also with cancers in her frontal sinus, throat, cortex of her brain, lungs, uterus and lymph glands. Trying to withstand the onslaught of two fatal illnesses at the same time for nine months became more than she could bear so she took her own life the night before her daughter's ninth birthday. Each year we celebrate dear Sherrie's birthday. I go through all the extreme agonies and pain I endured that fateful night.

I urge your Commission to make every effort, right now, to search out some ways and means to combat and cure this most feared and fatal disease.

CARL V. PAYNE
TURLOCK, CALIFORNIA

MARCH 6, 1977

I am taking this opportunity to relate to you my personal experience regarding Huntington's disease and the severe problems, financial, mental, physical, etc., that it inflicted upon my life and my family. It is my sincere hope that the information can be utilized and be of significant help to your Commission in making recommendations to Congress regarding the tremendous needs a family has when one or more members has this deplorable disease.

First of all, I will explain where the disease came from. My wife, Betty, inherited it from her father's side of the family. She passed away in August, 1976, a victim of Huntington's disease, a disease she had for nine years before she passed away at the age of 46.

During this nine years, it caused such mental conflicts on myself and my family, so that today we are in such an emotionally disturbed condition that it is doubtful if some will ever recover. It was also during this nine years that I lost my job with an aerospace firm and it is now my belief that due to the mental distress that I was under at the time, that helped bring on an early lay-off. This caused me severe problems in getting new employment at my middle age of life. I have had to accept a very low income type of employment, so as to provide a living for my family.

After my wife had this disease for about three years, it began to take its toll upon her physically in a tremendous way. The first years, it was more mental, and in the last years of her life, it was more physical. The very last three years of her life she became totally incapacitated and had to be confined to a skilled nursing home. I was not able to work, watch, and take care of her at the same time, and it was imperative that I work to provide a living. The financial expense at the nursing home destroyed me. I had to sell my home, and now I have nothing. I lost everything at the very last year of my wife's life. It was recommended by the Welfare Department that I obtain a certain type of divorce so as to allow her nursing home expense to be covered by Social Security of some type. This was very distressing to me and very soul searching, but that was what

it was going to come to when she died. Now what the future holds for my children, only time will tell, for this disease is hereditary and it is a distressing thought that this deplorable disease could still inflict even more suffering and mental distress in the life of my family. Also, in view of the enormous amount of other people suffering from this disease and in their future generations, I urge your Commission to make every effort to combat and cure it. Something needs to be done, something should be done, and I also urge your Commission to use this opportunity to voice the need for a national health insurance to the President and the Congress of the United States.

Los Angeles, California

April 15, 1977

RUTH PAYNE
LANCASTER, CALIFORNIA

MARCH 27, 1977

In regards to Huntington's disease, this illness is a very heartbreaking illness. I have seen my friend Ruth Krueger's illness getting progressively worse. It has caused marital, personal and family problems.

Ruth was one quarter short in getting her Social Security which I think she should have gotten. The financial needs are great for these people.

I think consideration should be taken in regards to homes for these people that are unable to take care of themselves.

It really upsets me to see babies born that have a background of this disease. There is enough suffering in this world.

I am a female, 51 years old, and married. I am working as a bookkeeper.

I hope this letter has helped in some way.

Los Angeles, California

April 15, 1977

MRS. KENNETH O. PENZES
MONROVIA, CALIFORNIA

MARCH 3, 1977

I feel a necessity and urgency to write in response to the hearing being scheduled to be held in Los Angeles on April 12, 1977.

The lives of my sister and her family have been drastically changed by Huntington's Disease. With no knowledge that the disease was inherent in the family line, the entire family went through a living hell during the time from the appearance of the first symptoms until the disease was finally diagnosed in her husband. The change in his disposition was unbelievable and the effects of that change were devastating.

My sister and her husband had four children, ranging in age from approximately 21 to 12, when it was discovered that he had H.D. The oldest girl completely separated herself from the family, married and had a baby, eventually committed herself to a mental institution and deserted her son (In the light of the fact that she herself is now a victim of H.D., this can be more easily understood.); the second girl suffered a nervous breakdown; the oldest son withdrew and became involved with drugs for a while; the youngest son seemed to suffer the least damage to his psyche at that point.

Through the long time it took for H.D. to run its course, crisis after crisis arose and there was nowhere to go for direction or help. The victims are not cooperative and so it becomes the obligation of the rest of the family to find their way out of financial, social, medical, and emotional problems. The need to establish some centers to help families thus afflicted is imperative since, in many ways, H.D. is more disastrous and debilitating than cancer and heart disease.

My brother-in-law died four years ago but since that time both his oldest son and daughter have shown symptoms of H.D. The same pattern of behavior is beginning to repeat itself in them. I fear not only for them but for the physical and mental health of my sister. The supportive services available through organizations who can offer information, subventions, and even possible medical assistance is absolutely necessary.

Another area that I feel needs attention is supportive counseling services to family members who do not as yet have any symptoms of H.D. and may never have them. My youngest nephew has had his goal set on being a doctor all of his life and has worked toward that aim. He is an excellent student and has worked for the finances involved in such an education. He

is now faced with problems in being accepted in medical school because he is a member of an H.D. family. He is attempting to lead as normal a life as possible but society will not allow it. He is being made to feel "guilty" about having a father who died of H.D. when the committee who interviewed him for entrance into medical school admitted that the reason for his not being accepted was H.D. Such barriers cannot be broken down in a short time but some measures need to be instituted to educate people, even people associated with the medical field.

The love we have for our sister and her family is no different than the love of most families who share the problem of H.D. Since detection of H.D. is often so very difficult, it is likely that the number of families thus affected is greater than now determined. I pray that we can work toward easing the burdens of all brothers and sisters thus afflicted.

Los Angeles, California

April 15, 1977

MARIAN M. PERKINS
FULLERTON, CALIFORNIA

FEBRUARY 28, 1977

I am a nurses' aide who has a patient with Huntington's chorea disease, and I don't know how to take care of her. This patient can't be left alone when up in a wheelchair as she thrashes around so much, and she can't talk so I have no way of communicating with her. Isn't there some way the Government could give a grant for research on this disease? We need to learn more about this type of patient so we can take care of them.

Los Angeles, California

April 15, 1977

J. W. PERSONS, D.D.S.
CAMARILLO, CALIFORNIA

FEBRUARY 23, 1977

In my practice I encourage all patients with muscular problems to have fixed bridgework replace missing teeth. Removable partial dentures to replace teeth are much more difficult to keep clean; they are hard to control in chewing; they trap and hold food against the anchor teeth they clasp around. Patients with poor muscle control have a harder time maintaining good oral hygiene and partials demand above average hygiene. Partial dentures demand dexterity putting them in and removing them. If there is a coughing spell or seizure they can be dangerous if swallowed or inhaled.

I would encourage use of an electric toothbrush instead of manual brushing.

I understand that choking to death on food is the most common death of Huntington's Disease patients. I would encourage preserving as many of the patients' teeth as possible to help them chew the food easier.

Gold castings as teeth need them will help support and save teeth that might otherwise be lost if simply repaired expediently with large silver restorations. The practice of patching large silver restorations is unsatisfactory because the filling or pieces of tooth can break off and be swallowed.

Los Angeles, California

April 15, 1977

BERNARD K. PETERSON
SAN LORENZO, CALIFORNIA

MARCH 4, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Being a member of a family heavily inflicted by this dreadful disease and experiencing first hand the problems connected with it causes me to make a personal appeal to you.

Los Angeles, California

April 15, 1977

SHARON PFEIFER
HAYWARD, CALIFORNIA

FEBRUARY 28, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

LARRY PIPPIN
WATERFORD, CALIFORNIA

FEBRUARY 26, 1977

As President of the Modesto Chapter, I would like to add my little effort to the cause. We are 42 to 50 members strong: afflicted, spouses, parents and those in sympathy.

We've encouraged each member to write you. We've been organized since December, 1976. Our local hospitals, doctors, Social Security, and Welfare representatives have been very cooperative in attending our meetings. I think we have learned much and getting very good response.

My wife, Helen, and I have an adopted son, 25 years old, afflicted with this dreaded disease. He is getting worse steadily. We first noticed his unusual actions for the last ten years. We have no way of knowing his family. A doctor diagnosed it as H.D. about four years ago. He will not cooperate with us to get a further follow-up. He thinks God will heal him. He is not belligerent at all otherwise. Goes to church with us regularly. Had to drop out of college because he couldn't keep up his grades. Can't communicate very well.

If you have any suggestions or can help us in any way, our chapter, please do.

Los Angeles, California

April 15, 1977

MRS. L. W. PIPPIN
WATERFORD, CALIFORNIA

FEBRUARY 25, 1977

We are the parents of a young man whose age is 25--six years ago his handicap was diagnosed as the dreaded Huntington's disease. He was adopted by us when he was three months old. He was a brilliant high school pupil and tried to carry on in college but H.D. has slowly taken over. We had no knowledge of this disease before he got it.

We all suffer from this disease, through watching and seeing him decline, and there is nothing we can do for him. I urge your Commission to make every effort to combat and find a cure.

Please help us so that help may come to Steve before he is just a vegetable.

Los Angeles, California

April 15, 1977

CAROLE PONCHETTI
FRESNO, CALIFORNIA

MARCH 22, 1977

I am friends with a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

AUGUST PROFUMO
SAN FRANCISCO, CALIFORNIA

JANUARY 2, 1977

I'm using this means to express how Huntington's disease affected my family and the difficulties that we felt because of this disease. My wife, Florence, is one of five children (living--she was one of seven), born in Portsmouth, New Hampshire on June 8, 1928. Two of her brothers passed away at the age of 2. Her mother died of an accident at the age of approximately 34. Two brothers and two sisters, the oldest sister, Mary, died of H.D., one of her brothers died of H.D., both at Napa State Hospital, and she died there also.

Currently one living sister and one living brother, the sister in a state hospital with H.D., so of the family of five, four have gotten the disease. Mary had six children and as far as I know they are all still living. Edward was married and had two children; my wife had two children; her other sister had two; the other brother never married. All of these living members are at risk with H.D. You can see the compounding of this disease and its consequences. Not all realize that they have the disease or possibly can get the disease. My boys are aware of it and so is one of her sisters--the other does not.

Florence and I were married in 1953 and we had our boys in 1954 and 1955. We were living in San Francisco and everything was going pretty normal until about 1959 or 1960--she was 31 at the time. The problems that I had were when she first had signs of mental depression, etc. I was not aware of Huntington's disease or anything like it, so at that time I tried to get some help through the mental health facilities in San Francisco. However, due to my financial abilities (I was a middle income family) my medical health plan was through Kaiser and it had no coverage for mental health or anything along that line. In trying to seek help in the area, it was almost impossible unless you were on welfare.

During 1961 and 1962 I contacted a local family physician who had delivered my first son and through him, he gave me at least some idea of what was happening. He thought that there was a possibility that it was a family inherited factor but he wasn't sure nor did he specify definitely. I finally in desperation returned to Kaiser and at that time they had developed an outpatient drop-in type of psychiatric treatment. I had expended funds for a private psychiatrist in San Francisco but then we stopped to go to the weekly visits at the outpatient clinic.

This was at Kaiser with Dr. Johnson of that staff. At that time the price was \$7.50 for a half-hour visit or \$15.00 a visit. Standard prices for psychiatrists at that time were \$25.00 to \$30.00 per hour. This expense continued through 1961 and 1962. She started to have more mental depressions and difficulties concerning her mental attitude and she also started to complain of weaknesses in the back and occasional headaches.

In 1963 she entered Presbyterian Hospital in San Francisco. Through our family physician we also contacted a Dr. (Irregetti) in that hospital (a psychiatrist) but he also did not diagnose what was really wrong with Florence. He decided after a period of psychiatric visits with Florence that she probably would need shock treatments which I finally agreed to through his insistence. So she had to go through these shock treatments in connection with her physical and mental problems. As you can realize, this was also an expense which was not covered by an insurance policy.

At the end of 1962 we were expecting a baby and the baby was stillborn; it had died the day before it was actually delivered. Shock treatments came after the stillborn--in the beginning of January. The shock treatments by Dr. Irregetti had resulted in an aborted baby; we were unaware that she was sick. We wanted to raise a larger family; I felt that the mental problems would eventually disappear. I was unaware of H.D. so we were trying to live our lives normally. She really tried to correct her problems--she gave it every effort possible. She worked when she thought it was necessary to keep her mind occupied; she would sing; she did everything you could possibly do for a person who was attempting to correct something she was unaware of happening to herself.

The shock treatments occurred the end of 1963, the same year we had the stillborn, and she had a miscarriage in 1964. Then during 1964 she returned to Kaiser; she lost faith in (Irregetti) and went to Dr. Johnson for awhile under the Kaiser Plan again. At the end of 1964 and the beginning of 1965, she actually entered the daytime clinic at Presbyterian and that lasted for about a month and for daytime visits from 9:00 to 5:00 it was \$90.00 per month, which was very reasonable. However, this came in addition to the hourly visits with Dr. (Irregetti) which was expensive. I was not a pauper at this stage, however, she started to have more and more mental problems and complained of the headaches from time to time.

In 1966 around April, she finally got so desperate that we went to San Mateo Public Health Hospital to receive some kind of medical attention. They couldn't recommend anything but they thought possibly maybe Agnew State Hospital--if she really felt desperate and thought she should be admitted--might accept her. Through her own recognizance, finally, she went there and was admitted. It was at this time that they diagnosed that she had Huntington's disease. The shock to me was tremendous. I went through a period of time where I probably could have become a patient in that institution as well; however, through a little calmness on my behalf or through my friends, I got through it.

She remained in the hospital at Agnew for about six weeks and, of course, there was an expense being incurred there also. I didn't have to contribute primarily because at the time my financial status was considered insufficient to handle the burden. Actually here it was six years, going to all the doctors, all the entrances to various institutions, and not one of them was able to diagnose the disease until she entered a state facility--Agnew State. The State Hospital has since then closed because of economics within the state. Reagan felt that state hospitals were no longer a necessity--that smaller institutions or smaller local groups would best facilitate the State of California. I think he was wrong.

She was let out of the hospital and she was now on drugs such as Valium, Librium, Thorazine, etc. We continued to try to solve the problem at home and she was pretty good for awhile. She was starting to lose her coordination, striking about a bit. She was once stopped by the police of Mountain View because they thought she was drunk. All of these stages are gradual. I can't describe specifically where she started with the uncoordination or the like. I do know that during certain periods of time she went through an awful lot of unusual activity with the idea of offsetting this illness. I mean she vacuumed for hours at a time, dust, move about, do things that were sort of unusual. She broke a lot of things because of her coordination; she spilled a lot of things and something along that line, which added to the difficulties.

In 1967 she was readmitted to Agnew for about two weeks; they already knew what the situation was but she was merely there for observation. They released her and she was brought home again; meanwhile she continued with treatment. In 1968, because of her mental depressions continuing, we started to use the facilities at San Mateo on a drop-in basis, which was primarily because she had been a patient at Agnew they allowed it.

This was at no cost to us--this was the only time I can recall receiving anything beneficial that didn't cost.

In 1969 she became very difficult, very depressed; she was just looking for some place where she could receive help. She desperately wanted help--she wanted to get well and kept insisting because of the boys. We went to San Mateo Hospital and talked to them to see how we could get some help. We went to Kaiser; we went to a welfare agency. No one could really offer anything that was within my financial range or even that I could qualify for. I couldn't qualify for Medical or any of the other so-called Federally financed projects. There just was nothing for this type of ailment. In desperation finally someone at Kaiser Hospital--they had a girl there that was experienced in welfare or some type of situation where she would handle the problem. I forgot what her title was. She finally arranged through Kaiser that we have a home care nurse come over at least twice a week to help me here. What I was trying to get at this time was some type of assistance where someone could come by, either on a daily basis or once a week, to help her--get her dressed, help her take a bath and just see to her needs. By this time she couldn't dress herself very well; she was able to feed herself but she couldn't prepare it. She had bathroom accidents and things like this.

During this one period of time, she went through the shower door so I could no longer allow her to use it. Finally we were able to get a home care unit, so during 1970, 1971 and 1972 the home care unit was taking care of Florence. She had someone coming in from the county at one time called the Sunshine Girl. They used to visit her once a week and just talk to her because she was still able to communicate quite well, even though she slurred a lot. Towards the end of 1971 and the beginning of '72 her speech was really slurring; she didn't know what she was saying. A woman came over twice a week and took care of her. I would dress her every morning before I went to work, leave some food for her in a room downstairs and make sure she was comfortable. When I came home in the evening I would change her clothes and put her to bed. This also included her monthly periods, etc., so for me it was very difficult.

Actually from 1965 and after I was actually maintaining the household myself--doing all the housework, cooking, and all the chores inside and out. She was no longer capable of doing much of this. Towards the end of 1972, she was able to walk around a little bit. On at least three of four occasions she got out of the house and was picked up twice by the police and taken to

the hospital. Then I knew I had another problem. I again contacted the county and told them I needed some type of assistance. They suggested again at this time that at Peninsula Hospital they had a doctor. At this time financing under Short-Doyle came into play--Short-Doyle is not assistance; it is just a method of financing whereby the individual will pay at a later date on a basis of financial ability. I arranged to get some type of financial agreement with them at the beginning of 1972 with the idea that she would still be at home and she would drop over to the mental hospital. However, around April of 1972, Florence got out of the house again, was picked up by the police and at this time I made arrangements with Dr. Fox, whom she had seen twice (and that cost also) to have her committed into the Peninsula Hospital in May of 1972 as a day patient. She made an attempt to leave the hospital there and at this time she was no longer capable of talking so that you could understand what she was saying. She just babbled and really started to become very irrational.

Anyway, I admitted her on a Sunday. I called on Monday and found out they had transported her to Napa State Hospital in Napa, California which is also a state facility. I had talked to Dr. Fox with the idea of trying to get her admitted into some local rest home and had also contacted the county. The county indicated their facility was at San Mateo where they could have admitted her was crowded, primarily because of the closing down of Agnew and they couldn't admit her because she would have to be admitted to a closed door facility. Now a closed door facility is for individuals who are mentally disabled and also they lock them up so they cannot get loose. Now I don't know whether this type of facility was necessary for Florence; I do know they could have had a facility locally where she could have been prevented from taking a little stroll because she wasn't that bad. The rest home facilities were explained to me at that time as ranging from \$400 to \$600 per month. I didn't have that kind of money so that was why ultimately she was shipped by ambulance at my expense to Napa State Hospital. She remained at Napa from May of 1972 until June 23, 1976 when she passed away.

She was given the best medical care that was possibly available in that hospital; however, it was a mental institution. In this particular hospital there are about 2 or 3 nurses, maybe one doctor, per ward; there are anywhere from 30, 40, to 70 patients in each ward, depending upon the availability of space; men and women are separated in different block houses.

The facility is fairly modern but it's still an old facility. It happened to be 90 miles away from my home so each Sunday I would visit her--a trip of 90 miles one way. It would take me an hour...I would spend about three or four hours every Sunday in this process. I would drive every Sunday morning up there, spend about an hour there, feed her, give her some candy and cookies, talk to her (even though she wasn't able to communicate) and then return home Sunday about noon. This I did every week during the entire four years that she was there. But each time I would try to have her returned back to San Francisco.

There was one aspect I didn't cover. In order for her to be admitted to Napa State Hospital in May of 1972, I was required to submit papers for divorce. We were divorced in November of that year, so technically I wasn't married to her after November of 1973; however, the expenditures that were incurred while she was in that hospital and due to the fact that she was in the hospital, it was required as a divorce settlement, to give to her or to her trustee--through the county--an \$8500 second deed of trust to my home. I was making payments of \$75.00 to \$150.00 per month initially; then I raised it to \$100 as payment on this second deed of trust. This continued on and is still continuing as of this day. Here she was located in a facility too far away for me to really visit her more frequently; my one son was able to visit with me on occasion, but still the distance was relatively far. She gradually began to dissipate in the hospital. I know they tried to give her as much care as possible, but near the end I'm not quite sure when a person appears to be of no hope, they sort of, I believe, they give attention but they can't really offer the patient any consolation or really give them enough food intake to have them at least be more comfortable. She was actually skin and bones when she passed away. She weighed 75-90 pounds. I believe this was mostly the illness although I'm not sure.

Recently they set up an estate for her when she passed away; she was receiving social security funds; I thought she was receiving MediCal but apparently she was not; there were some funds left to my sons either from the MediCal payments or the Social Security that was being placed in. Anyway, the expense of this hospitalization from May 1972 to June--four years and approximately one month or 49 months--was \$51,000; around \$1,000 per month. This is the bill that was presented by the State of California to the estate of my deceased wife.

There is approximately \$3,000 in the estate; I don't know where the funds came from. I think it is primarily my payments on the second deed of trust plus possibly from Social Security payments that wasn't mailed directly to the State of California. Of the \$51,000, \$7,000 came from sources probably from Social Security against the \$51,000 bill. There is currently a remainder of \$44,000 expense (hospitalization) which is a lien against my wife's estate. It will wipe out or dissipate the remaining funds that are in the account that would have gone to my sons. I'm not sure whether or not my \$8,000 remaining second deed of trust will also be part of state property and I'll be required to give them the \$8,000 instead of applying it as part of the liability. Overall it would appear that with all the anguish and suffering I've gone through and unrecorded expenses of all the past things that happened to her and the future expenses that I may incur, that this disease is by far a financial disaster in the overall picture of things. I know that I have spent pretty close to, in addition to the amount expended by the State of California or that bill, probably expended in the nature of \$10,000 to \$15,000 over the entire 16 or 18 years that this disease was in effect. However, I'm not quite sure exactly how much it would cost. I'm counting in terms of also items that have been subject to repair or a direct result of her illness where she had vitually broken things or something along that line.

Offering this testimony in the hopes that you'll find some solution to this particular problem--particularly in the terms of financing, there must be a facility available at least for this type of ailment. A closed door facility is not the solution. A mental institution is not the solution. A person who is old or a person who has this type of ailment should not be placed in a mental institution; it is not the proper facility for them. They are too crowded as it is. This illness should also be classified as a catastrophic illness as well, so at least individuals with H.D. or at risk could receive some type of funding. There is no insurance policy or coverage that an individual can get with this type of illness. You cannot get a life insurance policy; you cannot get a medical insurance policy--at least not at a low rate of insurance premiums. They are all catastrophically high.

It is hoped that this testimony will be of use to you and your Commission in making a determination as to what can be done to either alleviate this disease, find a cure for this disease and to provide some type of assistance financially or otherwise to

April 15, 1977

victims of H.D., both for those who have the disease and are at risk because at risk individuals can also incur liabilities in the future should they get it. I have that problem and I'm not looking forward to it.

In view of my past experience with H.D., I feel the following are needed:

1. Health insurance which will include coverage for H.D.
2. Life Insurance which will insure H.D. families (all members).
3. Home care nursing or aid during the daytime hours for working husbands or wives of H.D. victims. Child care aid if minor children are also involved.
4. A special care residential or rest home facility for the H.D. victim. (This is most important). The current treatment in mental health institutions is inadequate. Many are too crowded to give assistance or treatment to H.D. victims. I feel the same way about the treatment of the aged. A mental institution is not the place for the aged either.
5. Research--any kind.

Thank you for your attention.

Los Angeles, California

April 15, 1977

LOUISE S. RANDOLPH
ALBANY, CALIFORNIA

MARCH 6, 1977

I am at risk for H.D., 37 years old, married and the mother of a five-year-old girl who is, of course, also at risk. My great-grandfather, grandfather and mother died from the complications of H.D.

I remember my grandfather only dimly. He was a kind-faced man who shook a great deal and who walked as if he were permanently tipsy ("there was a crooked man who walked a crooked mile"). He died at Agnews State Hospital, strapped down in the psychiatric ward. He had been strapped down because they knew of no other way to control his outbursts of violence. Great-grandfather's outbursts of temper were dealt with by locking him in the closet in his own house.

More vivid and necessarily more disturbing to me are my recollections of the ways H.D. affected my mother. Like her father and grandfather, Momma had her flare-ups. Her anger was something to behold, and she alienated most of her friends and the larger family by her outbursts. Remembering her, I sometimes wonder if paranoia and anger are caused by the disease or by frustration at the shaking, twitching, peripheral vision and social ostracism. Another reason for her ever-decreasing circle of friends was that some of them, against all logic, believed her disease--like cancer--might be contagious.

She had trouble with coordination after about age 45. She took driving lessons three times in a row, but she never got her license. She simply could not get her eyes, feet and hands to work in pairs, much less to behave as if they belonged to one person rather than to three. Her coordination was impaired long before we were aware that something was seriously wrong. In fact, her erratic motor behavior and her outbursts of temper at this stage were attributed to menopause.

She was proud of being a bookworm, but the disease restricted her seeing to peripheral vision, so she had to give up reading; at the same time she was mostly confined by her condition to a chair or to bed. Unable to read, she "watched" TV for about ten hours a day--I suspect she heard more than she saw.

Her twitching and spasms were controlled by anti-spasmodics, and she was given Valium to keep her tranquil. On top of the tremendous drain on energy caused by spasms and anger, the disease affected her metabolism so drastically that her weight dropped from 115 to 88, then charged back up to 130 and finally plummeted to 88 pounds by the time she died. In the case of my grandfather, dehydration must have been one of the signs of metabolic change since my father recalls that he had demanded excessive amounts of water during his last two years.

My father kept my mother at home as long as possible, with a neighbor checking on her. He had to assume all of her responsibilities, a burden which left him with a profound respect for working wives.

A diagnosis of her disease might have come earlier than it did had she had not been as suspicious as she was of doctors. Leaving aside the legitimacy of her suspicions, there just is not a whole lot doctors can do for H.D. victims. Then, too, who knows how much of her suspicions were the product of her own fear of becoming like her father and grandfather?

Not the last of the unfortunate consequences of H.D. are the economic ones. My mother was penalized by the Social Security System, for although she had worked for ten years before she married, her employment had been in a period before Social Security became effective. She was also ineligible because she elected to be a homemaker instead of "working" and because she died before her 62nd birthday.

Besides isolation, money was the biggest problem for my father. The rest home costs would have left him with \$150 a month from his paycheck. Fortunately, his personnel manager, by doing some arm twisting, was able to get the costs paid from a medical insurance policy. Without such assistance, the financial consequences of H.D. would have been ruinous.

My biggest problems right now, since I have shown no symptoms, are fear and occasional attacks of despair. The despair was more common when my daughter was a baby. Now that she seems more capable of defending herself, I think about it less. Believe me, writing this letter is no help, either. Being at-risk is not the sort of problem for which confession provides catharsis.

What would help me? I would like a control for the disease SOON, something like L-dopa or insulin. Should I contract the disease, I want to be as normal as possible: to be able to go on knitting and reading, cooking and going to the bathroom, bathing and feeding myself just like I do now. In other words, I want to keep my self-respect and dignity. I desperately want this for my daughter too.

If such a control is not possible for me, please help to keep me from impoverishing my family. We are middle class, which means we pay taxes but get precious few benefits from them. My family would need in-home assistance and help for the cost of a rest home. Perhaps some rest homes do not have to be profit-making; maybe the state can help with a sliding scale of payments. Mine is the same request made by people burdened with the expenses of cancer treatment, care of the mentally retarded and the scores of other human afflictions for which our society provides little help.

My best wishes are with the Commission in its dedicated search for solutions to the problems of H.D. families.

Los Angeles, California

April 15, 1977

MARIAN RASMUSSEN
VILLA PARK, CALIFORNIA

MARCH 10, 1977

I have a dear neighbor who has Huntington's disease. When they moved here several years ago, Val Link was a very active, interesting and fun person. Gradually her health has deteriorated, until now she can hardly get around. When walking she shuffles along and she is not able to carry on a conversation any more.

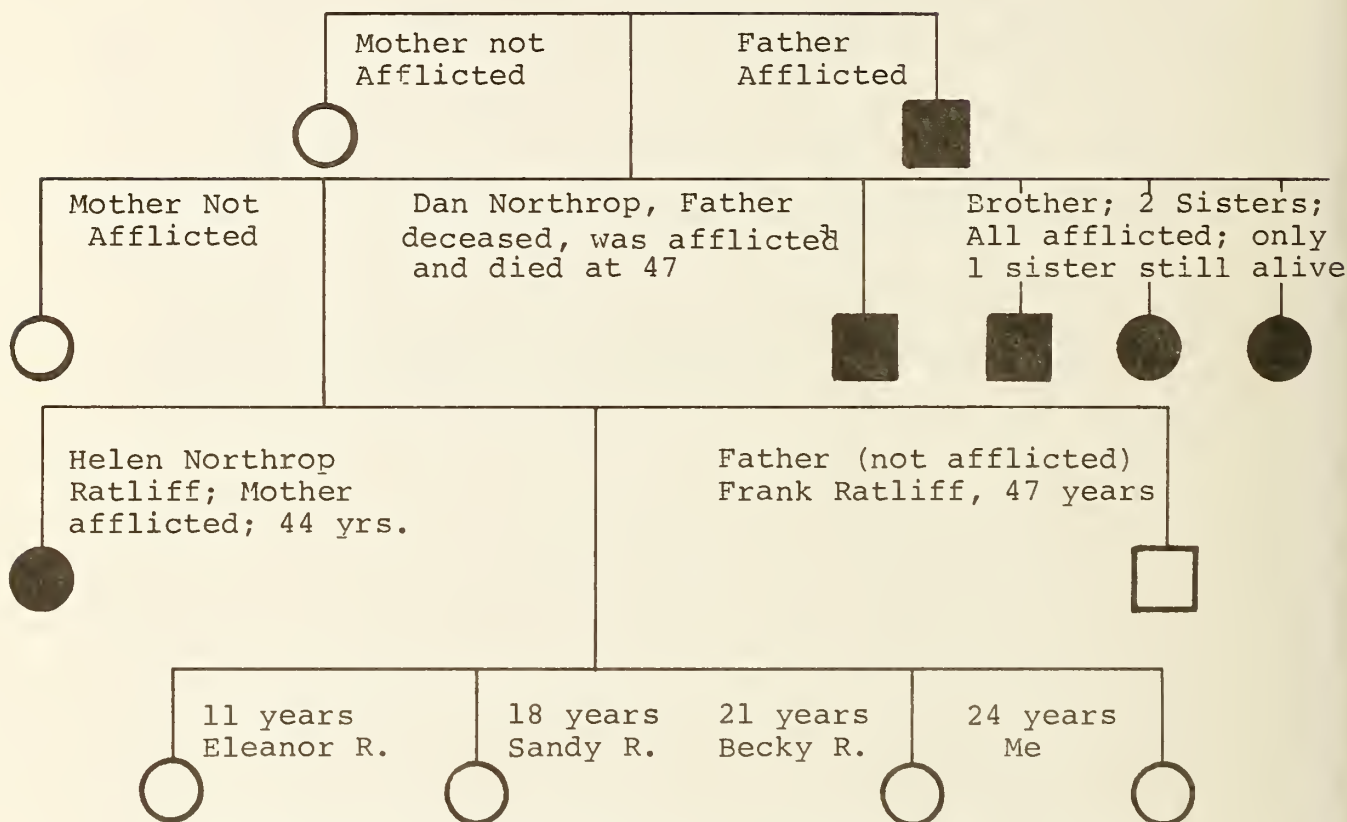
It saddens me to see her this way and not be able to help her. It is a terrific strain on her husband and children. Val cannot be alone, making necessary "live in" help, which is an added burden to the family.

My understanding is that not much is known about the disease or what to do for it. I feel strongly that there should be a concentrated study made now, to find medication or help for those with Huntington's disease. It may not be in time for Val, but it could be for her children.

MARGO RATLIFF
MODESTO, CALIFORNIA

DECEMBER 4, 1976

My name is Margo Ratliff. I am an "at risk" case. Here is my family tree.



I'm not a genetics expert so my drawing may not be right, but I think you can tell who all has it.

My mother's case is complicated because she previously had two nervous breakdowns and two strokes. She has difficulty speaking and continuously jerks her upper body and head. Her psychiatrist thinks that this is about her third year. The official diagnosis was made in November, 1975. She had claimed for years that she would get this but we kept taking her to doctors and they said, "No." She was "borderline schizophrenic," "neurotic," "high strung," "maybe epileptic," etc.

Finally she went into a hospital and had a brain scan done. While there she had a stroke. They evidently found out then that Huntington's had begun but did not tell my father or us--this was in November, 1973. I went with her two times to the Stockton State Hospital and asked the doctor this: "She claims she has Huntington's--is this possible?" They answered, "No, that is very rare--she is just nervous." They would give her a bottle of Valium.

At home she got worse and worse, became violent, hitting everyone, causing trouble with neighbors, having delusions about my dad having affairs and saying really insane things to people.

Finally after one particularly bad scene, my dad and I took her to the State Hospital to see if she could be placed in a psychiatric hospital. While we were there her doctor casually mentioned, "Oh, you did know that she has Huntington's." My dad and I were stunned. They knew this for two years (ever since she had had the brain scan) but somehow never bothered to tell us. This exact same doctor had told me twice that she didn't have it! I was very irate because I'm 24; I could have had a child by now, and I feel like they had a responsibility to inform me. If I would have had a child, I would be outraged.

Anyway, we kept Mom at home for a few more months, but the situation was really bad. Finally she agreed to go to the hospital for observation and there the family went as a unit and we talked to the psychiatrist and a social worker and agreed that to keep the family functioning, she would go to a boarding home and come visit once or twice a week. It was a very difficult decision. It was so sad that at one point even the psychiatrist's eyes watered and I thought he might cry too. (This psychiatrist was a different one than the earlier mentioned one.) She is now in a place where she does not have to cope with anything and there is someone there with her all day long. The family gets along better, the house is clean and runs smoothly, there are no more ugly scenes and when we are around Mom it's a loving and warm time. You can be patient with someone for an afternoon or a day but not every day.

A lot of people criticized us for doing that to our Mom. Even though that really hurt, I know that they just don't understand. They don't realize that it is also mean to have her stay at home by herself all day while we're at school or work. She leaves the stove on and burns her hands, stumbles and falls down on things, and chokes on her food.

I think that was the hardest part; we were all so torn up by the decision and then to get a bunch of criticism was just too much.

We just organized a local chapter last night and I was elected secretary. If you could send us some information we would be grateful. For it being such a rare disease we had 42 people show up at our first meeting. It's amazing too--all the different ideas people had about the disease. For our next meeting, we're going to get a good doctor to come and explain the basic disease because there is so much confusion about it.

If you need letters from the rest of my family, let me know. We'd be very happy to send you dates and names and whatever else you could use.

The last year has been such a nightmare but it's beginning to lessen now that I'm aware of how much is being done to fight this. I certainly don't want this disease for myself or my sisters, or anyone!

Thank you for your time.

Los Angeles, California

April 15, 1977

JOYCE REDMON
DENAIR, CALIFORNIA

FEBRUARY 14, 1977

I am an "At Risk" person facing the prospect of possibly inheriting the same disease that took my father at the age of 67. I have a brother, who at the present age of 45, has just lost his job because of this disease which he has inherited. I am 44 years of age and know that there is still a possibility that I can become afflicted with Huntington's at any time. But my main worry is that should I become afflicted, then our two daughters will face the future in fear of also inheriting this dreaded disease. I have several uncles (brothers of my father) who are in various stages of this disease. My father's father died at the age of 56, evidently with symptom's of this disease while his own brother had what was commonly referred to at that time as St. Vitus Dance (same movements as I see now in my brother). As a child I remember vividly the movements of this uncle. My father's father's mother (it is reported) had to be taken to a mental institution at one time until other children went after her to care for her themselves. This is as far back as I can trace this in our family. However, a daughter of my father's brother (my cousin) just died this past year at the age of 46 from Huntington's. My brother, it appears, is rapidly facing the final stages of this disease.

I would urge you to appropriate 99% of any monies that may be available into research for the cure and/or prevention of this dreaded Huntington's Disease. It is too late to help me now - but please give hope to our children. Surely our great scientist's can uncover the cause or can discover a cure. We put our hope in this.

Los Angeles, California

April 15, 1977

JO ANN REINHARDT
FRESNO, CALIFORNIA

MARCH 22, 1977

I am friends with a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

RANDALL H. REISWIG
REDDING, CALIFORNIA

MARCH 30, 1977

I have seen a man deteriorate from Huntington's disease and I am now witnessing symptoms of this illness develop in his children. It is a horrible affliction and I urge your Commission to do everything possible to combat it and cure it.

I would particularly like to urge support for research, as I believe that there are likely to be some spin-off benefits for people suffering from other illnesses.

MARY E. RICHMOND
REDONDO BEACH, CALIFORNIA

JANUARY 28, 1977

I attended a CCHD meeting in November in Los Angeles to hear Dr. Milton Wexler speak on H.D. and learn what was being accomplished on trying to eradicate this dreadful disease. Everyone was asked to write to the Commission telling of any experiences they have had involving victims of H.D. My son-in-law, Don H. Bradford, suffered with H.D. for 15 or 20 years without any of us ever knowing he had such an affliction, at least for most of that time. Also, none of us had ever heard of such a disease.

It was hard to understand his peculiar actions. He was kindness itself and acquired a larger and larger following from the students to whom he taught history at El Camino College, a community college with over 10,000 students in Torrance, California. Parents, as well as students, told me how much he had meant in their lives. He had a brilliant mind and retained his knowledge from once having stored it in his brain. His vision became bad. His mother began to read onto tapes so that he could keep abreast of his subject, which was Western Civilization. His death in 1975 was a greater loss to the community than one can imagine.

His wife, Mary Ellen Bradford, my daughter, has recently sent in a letter, and I would like mine to be added to hers, to tell this part of Don's death. She and her family had been asked to be sure and notify the Commission immediately upon his death, so that an autopsy could be performed and brain tissue be available for vital research. Mrs. Bradford, sensing death was imminent, called their family physician to set the wheels in motion. His death took place about 9:30 P.M. in a nursing home. After his death, we all returned to the family home. The phoning began in earnest. It was one of the most heart-breaking scenes I have ever witnessed and I hope no one else must experience. The doctors who are so concerned with H.D. and have offices in Los Angeles and who live there, could not be reached. Every laboratory called could not or would not send a technician to do the work. Even the Veterans Hospital would not help. It got later and later and the emphasis had been on haste. Finally, one man said he would come for \$500; and this noble family who were determined that Don's death was not to be for naught. The greatest gift from him was to be tissue that might save the many victims and his children. The

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family accepted the responsibility for raising the \$500.00. They also had to locate a mortuary where the work could be done. It was though the whole world had turned its back on this family, trying to do a great good for humanity.

I hope my accounting of this experience will help emphasize the need for well ordered plans for success of all those needing assistance in the future.

With a full heart and sincere good wishes for the success of all those working so faithfully in this cause, I am

Mary Richmond,

Mother of Mary Ellen Bradford

Mother-in-law of Don Bradford

Grandmother of: Judy Bradford Rini (at risk)
Dulcy Bradford Rimac (at risk)
Andrew H. Bradford (at risk)

Great Grandmother: Melora Rini
Christy Rini
Nicole Rimac
Peter Rimac

Friend of Ethel Bradford, mother of Don H. Bradford

Friend of June Bradford Clark, sister of Don H. Bradford,
an H.D. victim and mother of Amy and Allen Clark

Los Angeles, California

April 15, 1977

DULCY BRADFORD RIMAC
VALENCIA, CALIFORNIA

FEBRUARY 28, 1976

I would like to think that the only ways I have been affected by being at risk are positive ones, but that would be a bit short-sighted. My plans for every phase of my life are affected by my being at risk. No part has been forgotten. All preparations must be completed, all decisions made, everything must peak when I turn 39. That is the age at which my father's deterioration began to show, and the age at which I have good reason to expect the same.

Being at risk has helped to keep my life and plans in a sharper focus than would be true for most people. I have a carefully laid out timetable for the next 10-15-20 years. And I am following it. I do not find it debilitating to be at risk. I have found it to be a very motivating factor in my life. But I would add that I am unusually lucky in having a large family who provide a loving and supportive atmosphere.

It is difficult to face the possibility of such a debilitating disease striking me or one of my siblings; but how much more difficult to be struck and not know what is happening. How horrible to begin the process of degeneration without the slightest idea of the cause, the reason. Such was the situation that my father faced. And as he was frightened and puzzled by what was happening, my siblings and I were outraged at his behavior--unable as we were to grasp what was taking place. Diagnosis was difficult, in that although my father knew something was wrong he tended to deny what was happening to the doctors. Some aspects of the degeneration he could talk about and others he would deny. After the diagnosis there was generally a strong feeling of relief for all of us in the family; a sense that at least we knew what "it" was and we could get a handle on it.

The first neurologist my father saw, the one who diagnosed H.D. as the problem, was correct in his diagnosis but wrong in most everything else. This prominent West Los Angeles doctor seemed curiously uninformed about H.D. Among other things, this neurologist had never heard of CCHD or Woody Guthrie. It was my sister who remembered hearing of a concert to benefit CCHD, and she provided this doctor with information for future

referrals. Not long after the diagnosis was made, my sister and I were visiting our family physician seeking advice on managing family relations. We were concerned about "our place" in the care of my father. The physician was most kind and understanding about our concerns and reassured us. After that part of our visit was over he began to discuss our father's recent diagnosis. He was interested and as pleased as we were that finally there was a diagnosis that seemed to fit. He then proceeded to tell my sister and I that we had no cause for concern because we were women and H.D. affected only men. It sounded like a great idea, except for one nagging flaw. That flaw was the inescapable fact that whatever my father had in the way of diseases, his older sister obviously had it too. Her condition physically seemed to parallel my father's, but emotionally she was much worse. Her personality seemed to be deteriorating.

Sometime later the noted neurologist had the whole family, excluding my father, come to his office for a consultation session. Included in the visit were my mother, my sister and her husband, myself and my husband, and my brother (at that time not married). This doctor explained to us all that he knew of H.D. and what we all could expect for the future, both my father's future and our own. He was accurate, as far as I know, in the information he gave us. He affirmed what we already knew. His session with us was very brief and very cold. In conclusion he told us three siblings that we ought to seriously consider sterilization and that having children for us would be very unwise. At the time I simply nodded my head in solemn agreement. I came away from that session feeling rather glum, and it took a long time before I realized just why. I appreciate the fact that H.D. is not an easy thing to deal with, no matter how pleasantly one tries to approach it; when you are handing it out as a diagnosis somebody's life is being completely reshaped. I basically knew what was coming before the session. We all did. It shouldn't have hit me hard and yet it did; why? I think because he handed it down to us as if it were some "graven tablets." He told us the facts; he laid out the timeline for us all. He told us all we ought to consider sterilization and that was all. He offered no suggestions as to how we could handle the situation other than his remarks concerning children. What we needed were counseling referrals, information about CCHD, and perhaps a little compassion. That was all 6 years ago and we still have not had a counseling session, but in my family there is a free flow of ideas and talk about the situation and at this point we are all handling it very well.

My father spent 5 years at home after he had to retire. The first 3 years of that time were very difficult ones for him. He was very depressed and moody a good deal of the time. It took quite a while before he became adjusted to the life at home after leaving a stimulating career. My mother worked full time and so was gone during the daytime, so my father's mother moved in with my parents. Although she was 76 at that time, she was still strong and healthy enough to be able to take care of my father, and this she did do ably for the next 5 years. His depression and withdrawal continued until it reached an absolute bottom in spring 1973, when he tried to commit suicide. It was a very clumsy attempt--he was not hurt--but nonetheless a genuine call for some help. It was at this time that my mother got the name of a new doctor who was treating already a couple of H.D. patients and with some good success. The doctor began by changing my father's medication, saying the medicine he had been taking would probably have made him very depressed and quiet. There was a marked change in my father caused not only by the medication but also by this doctor's attitude toward and treatment of my father. He was open and interested. For the next 2 years my father's condition stabilized; he was at peace with himself, with his condition, and with his family. By the end of July 1975 his condition had deteriorated to a point where my mother and grandmother agreed that they two could not any longer manage his care. My mother visited several convalescent hospitals in her area, and found them all lacking. Finally her physician recommended a place that she found adequate, and so he went.

The biggest problem with the convalescent hospital for my mother was accepting the fact that it had to be. She went about it very carefully, selecting one that was pleasant enough. It hurt me, too, to think he had to go in; but I also knew that I could offer nothing better in the way of a solution. I had two small children to care for. My father fortunately was only there for approximately 6 weeks. He died there on September 10, 1975.

For myself, being at risk has spurred me on to do some very careful planning for the future. We have gambled and have had two children--we will have no more. Neither my husband nor I could imagine life without a couple of kids--we both come from strongly knit families. I was young enough when they were born that they should both be teenagers before I begin to show any symptoms.

April 15, 1977

In the next 3-4 years I would like to go back to working outside the home. I have an elementary teaching credential now, but I realize that if I want to work as long as possible I would be better off with some kind of job that doesn't require the physical stamina that an elementary school teacher must possess. And so, at this time, I have started back to school part time to begin preparations for a second career.

My husband and I are carefully planning what our financial situation will be in the next few years. Our children already are well provided for. There will be money for them no matter what happens to either of us. We recently were shopping for life insurance and the Aetna Life Insurance Company was very eager to insure my husband but turned me down. I have since found other companies that will insure me, but there is no sense in giving them Don to insure if they wouldn't take me, too.

I hope that I have covered all aspects of the disease that you are concerned with. If you have any questions of me I will be happy to testify.

JUDITH BRADFORD RINI
HUNTINGTON BEACH, CALIFORNIA

MARCH 26, 1977

This letter is a response to the request for letters detailing experiences with H.D. I find this not only overwhelming, but impossible. Words cannot describe the tragic, shattering results of this disease.

I must also add, at the outset, that my family's experiences with H.D. have not been as devastating as the experiences of other H.D. families I have met. We face H.D. with as much positive force as possible. It is the only way to survive under the circumstances. This is a vulnerable rationalization.

A year and a half ago my 53-year-old father finally died of H.D. He spent his last month in a very unsatisfactory nursing home. He weighed approximately 100 pounds; he was 6'4" tall. His death marked the end of a 19-year struggle, not just with death, but worse, with the cruelty, stupidity, and ignorance of our society.

His struggle began when he was born and inherited an H.D. gene. Due to separation, lack of communication, and what must be supposed incorrect diagnosis, no one knew my grandfather died of H.D. My father never suspected the genetic time bomb he carried.

When he was 18 years old, he expected a lot from life. Dreaming of being a poet and/or a journalist, he left a small town and journeyed to UCLA. His education was interrupted by marriage, World War II (he was a fighter pilot in the Navy), and my birth. When my sister was born, my father was teaching in a high school. By the time my brother came along, my father was a professor at El Camino College.

When my father was 32 years old (I was 11), he was a handsome, brilliant, loving man, and an excellent and popular professor. He had become an adopted father to all the neighborhood children.

And then, very slowly, it began: the clumsiness, the absent-mindedness, strange physical complaints. From then on some mysterious force began to pull and warp everything in our family. My brother was only about four years old at the time.

While I was in high school the tension began to mount. My father saw every kind of a doctor one can imagine. He was insulted and degraded. This once proud man was ridiculed for his eccentricities.

The horror remains his loss of dignity and our anger. As my mother tried to protect him, we children fumed. "Why wouldn't she admit he was crazy?" A terrible change in character had occurred, not only in my father, but in our whole family.

The ultimate humiliation for my father happened when I was in college, when he came to me and cried, and explained that Mother would have to go to get a job; he didn't feel he could keep working. Fortunately, my mother had just finished getting her teaching credential. She had raced through school, in a panic, knowing her world was disintegrating.

My father's professional career was ended when he was hit by a car while bicycling (probably due to his lack of coordination). This accident resulted in two brain surgeries in a week for blood clots and a bout with infectious hepatitis a month later. It was a miracle he survived.

Still searching for an explanation for symptoms he went to yet another neurologist. He was finally diagnosed as an H.D. patient. (I diagnosed H.D. before the doctors did! I had read about it and I knew my aunt had it too. Our M.D. had said, "No one gets that--too rare. Only men get it," etc.)

The neurologist called all the family together (excluding my father) including: my newly-wed sister, my 17-year-old brother, my mother, and me (I had just had my first baby). He told us, in as cruel and antiseptic a manner as possible, that we should be sterilized as quickly as possible. Any other solution, he made perfectly clear, would be asinine and ignorant. I asked him about an organization that could give us information, genetic counseling, etc. He said there was no organization. No one gets H.D.

His prescription for my father was a heavy tranquilizer. So for two years my once proud father drooled and nodded. He lay in bed. He never dressed. A rest home seemed the only answer.

He didn't know he had H.D. My mother was afraid this information would destroy him. She told him he had suffered some brain damage from his accident. His friends stopped coming around.

His ultimate answer was attempted suicide. In his frail condition, he managed to place a rickety ladder up against the house. He climbed up and jumped off. He survived with only bruises. His reaction, "I can't even kill myself."

All this time, the family members were being torn apart. Tension and despair separated us. Finally I found CCHD from an article in the paper. Through CCHD we found Dr. Salick.

My father, in essence, was reborn. Salick told him of H.D., but gave him hope of future cure. He administered more appropriate medication. My father dressed, walked, laughed, and loved again. Salick's secret was to treat my father like a man, not a freak. Not such an amazing medical feat. My father, as a result, approached death an innocent, spiritual, loving child.

In the meantime, my sister, brother and I developed our positive philosophy towards H.D. that it's better to live a short, quality life, sharing love (as my father had) than to live as many "normal" people do. We also began to live for one day at a time. These are fragile rationalizations indeed.

CCHD also gave us hope for a cure, to the point that I don't worry about my two daughters or my niece and nephew getting H.D. I have faith in you, our society. Faith that you will finance research, educate society, help financially devastated families, set up hospitals for diagnosis and treatment, and special care facilities.

However, every day of my life there is a shadow present. Time has speeded up, and I am 31 years old. Help me.

I am willing to testify if necessary.

Los Angeles, California

April 15, 1977

MRS. WILLIAM ROSEWALL
TURLOCK, CALIFORNIA

MARCH 3, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

This is a very cruel disease. I lived with it for over five years, slowly observing my husband deteriorate mentally. He is a 64-year-old man and now acts like a 5-year-old child. I am trying to care for him. I thank the Lord he is able to be up but we are just existing. I, too, must suffer with him as I'm not free to go anywhere without him. Our four daughters, now married, are at-risk and so are our ten lovely grandchildren.

Please help us. We would like to live and serve too.

Los Angeles, California

April 15, 1977

MAXINE W. SCHEIBNER
EL CAJON, CALIFORNIA

FEBRUARY 19, 1977

As a mother of an adopted son, at risk with Huntington's disease, I urge you to act upon our pleas for help from this dread disease. A special plea toward research funds hoping for a treatment or medication that will minimize or relieve the physical symptoms of this disease.

The impact of H.D. was perhaps a big factor in my son's wife weeking divorce. When they came face to face with the possibility of the disease, it seemed too devastating for her to face, even though she knew my son was an at-risk person before the marriage.

Our big problem with my son is that he doesn't care to discuss his fears with us. He is a heavy drinker--even at 23--though I do not know if this is his inner worry about his future possible problem or not.

Please encourage more research and more information about H.D. to the public.

Los Angeles, California

April 15, 1977

MRS. H. J. SCHNEIDER
EL CERRITO, CALIFORNIA

MARCH 30, 1977

I have just heard of your Commission and want to express my extreme desire that you do all in your power to combat and cure this dreadful disease.

My father was in a hospital seven years with it, and disabled many years before that at home. My sister was the same story and my three other sisters and I lived with the fear of getting it. Now I see a niece and nephew living with the same fear. We didn't know then what the disease was, or that it was hereditary, or none of us would have had families. Please do all you can.

Los Angeles, California

April 15, 1977

FRANKLIN J. SCHURR
ORINDA, CALIFORNIA

MARCH 1, 1977

May I add my thoughts to the testimony that you are taking relative to the need for fighting against Huntington's disease.

My wife was first diagnosed as a Huntington's disease victim in 1963. At that time we had four children, ranging in age from 5 to 14. While there had been problems with my wife's father, these were not known to her as an hereditary disease.

The four children are in their early working years, or in college and, in general, are doing as well as can be expected with the health hazard that they face. They do not talk about it much, but the cloud of Huntington's disease must surely affect their long-range goals and motivation.

The Committee to Combat Huntington's Disease, headed by Marjorie Guthrie, has accomplished so much in the 14 years since this disease was first diagnosed in my wife. The Committee's approach has brought into the open the large number of families that are involved; and through the Committee's efforts, a substantial number of medical people have been alerted to the scope of the problem.

In 1963 and 1964, when I tried to determine what research and study might be going on, I could find no work of any substance.

Now there are many study projects, approaching the problem from many different angles--many dedicated people trying to find prevention and cure.

The Committee to Combat Huntington's Disease does not have the financial strength to support these activities. They must enlist and encourage action by groups who have means to research funding.

So much is being accomplished by many small groups working at medical research centers. But it appears to me that their work could be strengthened and coordinated by an overall Research Planning Unit--a body that could direct certain

research groups into areas that would lead on from work accomplished by others; that could be a clearinghouse for exchange of information; in other words, a basic coordinating body to get the best use of the experience and talent of the various groups involved.

Then there is the question of funding the research work.

It is amazing how much progress has been made in the last 14 years. The cure or prevention is not here yet, but the momentum is there; the problem can be more closely defined; there is hope for the young people such as my children.

However the funding may be handled--perhaps through a Research Planning Unit such as I have suggested--let's not lose the momentum that has been generated.

As more areas of possible solutions are studied, the funding needs will be surely greater. As research proceeds into more complicated organic chemistry, the costs of the advanced techniques will be surely greater.

Let's be sure that none of the work is curtailed by lack of funding.

There are just too many young people at-risk!

We must keep up the work that was started by Marjorie Guthrie and supported by so many dedicated people.

Los Angeles, California

April 15, 1977

MRS. MILTON SEIGEL
LOS ANGELES, CALIFORNIA

FEBRUARY 18, 1977

In our family we have Huntington's disease.

We need experience in obtaining a correct diagnosis. The doctors need help in helping us. They just shrug their shoulders and say there is nothing to be done.

We need new medicine to help us. Also we have children who are scared to death of what may happen. They had children before we knew about my husband.

Whatever you do will be a big help because we hardly have any!

Thanks.

Los Angeles, California

April 15, 1977

LORRIL SENEFELD
FULLERTON, CALIFORNIA

MARCH 25, 1977

Huntington's disease has been described as a time bomb which may or may not go off in individuals in afflicted families. The anguish this causes cannot be described! The fact that it usually shows up at mid-life is heartbreaking. Then one knows that one's children or grandchildren may be at risk. One lives with that sick dread every hour.

My husband's sister has just died after being ill for almost ten years. Her death was a blessing because of the deplorable condition she had degenerated to. She left four fine young adult children and a husband who had aged beyond his years.

H.D. is such a humiliating disease. It robs a human being of all dignity.

My husband seems to have been spared and yet that cloud of fear is something I live with. It is such a frightening thing that we do not talk about it.

I am so grateful that your Commission has been formed and that research is going on.

Los Angeles, California

April 15, 1977

SUSAN SENEFELD
SACRAMENTO, CALIFORNIA

MARCH 12, 1977

My aunt has just died of Huntington's disease after years of slow deterioration. My cousins are all "at-risk" and must plan their lives knowing they may possibly have this crippling disease. Most likely they will not have children for fear of passing it on.

I urge the Commission to do everything possible to obtain funds for research. We need to stop the suffering of victims, their families and friends. Finding a cure for Huntington's disease is of the highest priority.

Los Angeles, California

April 15, 1977

ESTHER SHINGLEDECKER
HAYWARD, CALIFORNIA

MARCH 1, 1977

I am the spouse of an H.D. victim. My husband passed away ten years ago. However, we went through an almost unbearable ten years.

I am sure there are other diseases as bad but when it is you going through it, you don't think so.

Since we had gone through the same experience with my husband's mother and his older sister, he realized what he was going through most of the time. The terrible part is you know it isn't over, ever. It is inflicted on every generation.

We have a daughter and now a grandson. I pray every night there will be some cure or at least some relief for this terrible disease by the time they develop H.D. if they are going to. My daughter is now 41, so is just going into the dangerous age for it to develop.

Please help us to combat and cure this terrible disease.

Thank you so much for your every effort.

Los Angeles, California

April 15, 1977

MARILYN R. SIMONS
BRENTWOOD, CALIFORNIA

MARCH 21, 1977

My mother has Huntington's disease. I find the multiplied pain of dealing with my mother's illness, the risk to myself and my children, and the risk to my brothers and sister overwhelming.

I have started to write this letter several times. Each time, I have stopped. Each time I have been blocked by the absurdity of the necessity to write a letter explaining what those of us who live with H.D. need, when to me it is so obvious.

Although the needs are many, the highest priority is research. Therapy and the education of both doctors and the public are also necessary, but none of that would be necessary if a cure for H.D. were discovered.

I can't tell you how I hope that one day we will be free of living under the tremendous pressure anyone who lives with H.D. feels. I do hope more funds are made available for research.

Los Angeles, California

April 15, 1977

JEAN SLADE
LOS ALTOS, CALIFORNIA

NOVEMBER 20, 1976

This opportunity of expressing myself as an "at risk" H.D. person is a rare one indeed! I want to thank everyone responsible for that one chance!

It is difficult to send my mind reeling back through a host of vivid memories, trying to decide which ones will trigger the minds of others towards a disease which has been medically diagnosed for more than one hundred years and continues to take its toll in the daily lives of all of us who are genetically recipient of Huntington's disease. We do need help and hope; and if my related experiences to the Commission give support to either of these factors, then I have succeeded in some small way.

I am one member in a family of seven children. Twelve years ago, my mother was diagnosed as having Huntington's chorea, "A disease," I was told, "which would be fatal not only to her, but also to approximately half of her children," which meant three or four of us. There are thirteen grandchildren to face the fact, in turn, that they would have a 50/50 chance of inheriting H.D. if one of their parents developed this disease. "And furthermore, there was no way of detecting H.D. until the victim had reached somewhere between the ages of 35 and 40 years." These facts were presented to my father and myself at a hospital that was 2,000 miles away from where we lived. I was left to inform the family as best I could because no one knew any more about H.D. than I did, including an array of medical people with whom I made immediate contact. Looking back at those twelve years could be summarized by stating that: (1) my father has cared for my mother without any help because there was none; (2) my family has suffered severely because there was no one with whom to talk concerning H.D.; (3) one of my brothers fought his fear for seven years and sought medical help throughout that period of time, and when he announced to his doctor that he was going to commit suicide, the doctor told, "Go ahead--you don't have the guts to do it." Ah, my friends, he DID DO IT! HE SHOT HIMSELF THROUGH THE HEAD!

After that happened, I made a promise to myself that we had to find help fast or this nightmare could occur again and again for who knows how long! And so I involved myself with

the creation of an H.D. chapter nearby. I spent all summer working to put it together--newspaper articles, TV and radio shows, postering, meetings, working with medical people, visiting with H.D. families, etc. I learned a lot about H.D. through my work, people were generally excited to become involved in helping other people who were less fortunate than themselves. I felt really useful doing what I could do to provide some hope--"hope?" There was no hope! There was information about so many kinds of projects in research of H.D., but our family was still basically where they were 12 years ago! There were new drugs to help control H.D. victims, but who wants to marry a pill for the rest of his life? There were drawers of H.D. files, tracing this family and that--but that's all it was--just a file! There was nothing being done to help all those people in the record file and the doctors weren't being honest with some of the families when they knew someone else had developed H.D.--it's still not handled without secrecy!

Then the time came when my mother couldn't be cared for by my father any longer, and we tried to have her admitted to a nursing home nearby so that he could visit with her. Since we could not afford a nursing home for her, we applied for assistance. The only place the government agencies would allow her to go was a MENTAL HOSPITAL! And we watched her with hollow eyes as she spent six months in the geriatrics ward of a mental institution, 105 miles away from her family and her home. Can you imagine what that did to all of us? Can you imagine what it did to a lady who has all her wits about her (better than I do) and is being punished because she can't walk by herself or feed herself or speak clearly? My father took her home again and tried to struggle on by himself. At least he and my mother have regained some measure of dignity.

Last summer I was able to convince the necessary authorities to allow my mother to be admitted to a hospital ten miles from her home. What a blessing! My father can visit with her as often as he wishes. The nurses know my family and do the best they can for her, but she often is black and blue from her elbows down to her hands because there are no beds with a protective padding for H.D. patients.

There is no special hospital equipment designed especially for the care of H.D. patients such as wheelchairs that give head and neck support for eating and sitting up, or cups that don't spill liquids, or equipment designed for washing their hair. My mother used to like to go for a ride in the car. Why aren't there special seats designed to support an H.D. passenger?

Perhaps we should have a catalog of equipment suited to the unique needs of H.D. patients and their care. This equipment could be available for rental through your local H.D. chapter or the Red Cross. It should be free to H.D. victims!

I have a fantastic dream of an H.D. Center with accommodations for all the needs of afflicted families. This could include a hospital for bedridden patients which would provide not only recreational activities, but also physical therapy. The medical staff would be hand-picked because of their knowledge and interest in H.D. A team of doctors (geneticist, psychiatrist, neurologist, researcher in H.D.) would provide the necessary guidance for the nursing staff and the voluntary workers (which would include H.D. family members). There should certainly be a program of counseling provided for all H.D. family members. This H.D. Center could be the headquarters for the local chapter and all related activities could be conducted through this organization.

In order to transfer this dream into reality, it is necessary to find financial support. Millions of people have willingly contributed their time, money and their creativity in order to show others that we need help in combating this disease. The emphasis has been placed on research because we have not won the battle until there is a cure for H.D. and only research can produce this result.

I am truly grateful to this Commission for its contribution concerning H.D. and hope that your conclusive evidence will lead toward the necessary support in finding a cure for this disease.

April 15, 1977

CLAUDIA SMITH
TORRANCE, CALIFORNIA

FEBRUARY 14, 1977

My name is Claudia Smith. I am 39 years old. I am "AT RISK". In my family, Huntingtons Disease can be traced back to my great grandfather. Next were my grandmother, mother, older sister and brother. The illness was not diagnosed correctly until a few years ago. My sister's doctor was the first to say H.D.

The research to find a cure for H.D. is of most importance. Running a very close second would be a test to determine if a person, at risk, will or will not inherit H.D.

The not knowing is a very traumatic thing. Things that are meaningless to most people, can be frightening to someone at risk. Little things like losing your balance for just a moment, bumping into something, stumbling or even trouble pronouncing a word, are intensified a hundred times. To us, this could be the beginning of H.D. I might add that all of the above symptoms give the impression of drunkenness. I know of two incidents where an H.D. patient was stopped by police.

Because of the late onset of the disease, you have many years of worry about yourselves and your loved ones. You must make many difficult decisions. You must decide on marriage, having children of your own and whether or not to inform friends of the disease and heredity factor. It may be unconscious but people do tend to stay away from the H.D. victim and family.

The financial drain of the family is tremendous. If you're lucky enough to receive medical, at least, you can stay in a relatively nice convalescent hospital. Even then many items and medicine must be paid for by the family.

The emotional drain is even harder on the husband or wife and the other family members. The personality changes that come with H.D. are difficult to understand and cope with. Some of the misunderstandings could possibly be helped by an early diagnosis.

My family and I try to help with all of the research projects that we can. We have all taken advantage of talking to a genetic counselor working with CCHD. There should be more of these people available to us. Sometimes you just need someone to listen.

For myself, my two children, my brother, my nephew and three nieces, all of whom are AT RISK, I thank you. Please don't waste precious time debating. Our time may be limited. We need help now!

Los Angeles, California

April 15, 1977

MRS. JAMES E. SMITH
NORTH HIGHLAND, CALIFORNIA

MARCH 3, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, my husband in particular, the families, and the future generation of our family and other afflicted families, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

MRS. ROGER E. SMITH
DENAIR, CALIFORNIA

MARCH 21, 1977

My father has H.D. and my mother and I are suffering with him just in trying to care for him. With Jesus Christ living within me, I have peace even in knowing the chances I have of having the disease myself. But that doesn't mean I am not very concerned about my own two precious daughters, or my three other sisters and their families that may also have it.

Our biggest problem now is to help my mom keep her own sanity as she cares for him. He is worse than caring for a 3-year-old child. He only sleeps short naps and never lets my mom sleep at all. He calls her "Ma-Ma" and won't let her out of his sight. I have to care for him some to give Mom a rest, but then he is crying and asking for her the whole time she is gone and this makes him even more up-tight.

In view of the enormous suffering H.D. inflicts on my dad and other victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

BETTY SMYTHE
FRESNO, CALIFORNIA

MARCH 22, 1977

I am a friend of a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

PHILIP R. SNOW, D.D.S.
CAMARILLO, CALIFORNIA

MARCH 3, 1977

In my practice I encourage all patients with muscular problems to have fixed bridgework replace missing teeth. Removable partial dentures are much more difficult to keep clean; they are hard to control in chewing; they trap and hold food against the anchor teeth they clasp around. Patients with poor muscle control have a harder time maintaining good oral hygiene and partials demand above average hygiene. Partial demand dexterity putting them in and removing them. If there is a coughing spell or seizure they can be dangerous if swallowed or inhaled.

I would encourage use of an electric toothbrush instead of manual brushing.

I understand that choking to death on food is the most common death of Huntington's Disease patients. I would encourage preserving as many of the patients' teeth as possible to help them chew their food easier.

Gold castings will help support and save teeth that might otherwise be lost if simply repaired expediently with large silver restorations. The prac-

tice of patching large silver restorations is unsatisfactory because the filling or pieces of tooth can break off and be swallowed.

I usually refer patients with muscular control problems to a local practitioner who uses nitrous oxide (gas) when working. The nitrous oxide plus a local anesthetic sedates the patient enough so that the expense of a general anesthetic and hospitalization for dental care is avoided.

Los Angeles, California

April 15, 1977

BILL SPARKE
CLOVIS, CALIFORNIA

MARCH 22, 1977

I am friends with a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

DEANNA SPENCER
CLOVIS, CALIFORNIA

MARCH 21, 1977

I am a very close friend of an H.D. family.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

SHIRLEY SPRAGUE
FRESNO, CALIFORNIA

MARCH 22, 1977

I am friends with a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

BLANCHE POINDEXTER SQUIRE
BERKELEY, CALIFORNIA

MARCH 24, 1977

Up until recently, for nearly two years, I cared for a woman in her fifties who was afflicted with Huntington's disease. These were the last two years of her life, and I truly could write a book telling of the day-to-day experiences of attempting to care for such a patient: the tears, the prayers, the determination.

In as few words as possible, I should like to mention some important points, at least I found them important, as I worked with the patient. My experience has taught me families desperately need HELP to care for their afflicted member(s), they need HELP to save themselves from breakdown, and HELP to combat the disease.

IMPORTANT CONSIDERATIONS:

1. Emotional strain on the caretakers, i.e., nurse, family. Really, this is beyond describing. The disease is the most dreadful one can imagine. The patient's psychotic behavior forced the long use of strong medication which produced undesirable side effects.

2. Physical strain on the caretakers. I came to a point of physical and emotional breakdown. I was able to continue caring for the patient five days a week (relieved usually by the husband evenings and weekends) for several months until I was forced to find a person to relieve me on Mondays. Four days became my limit. I suffered injuries to my back and feet (the patient's weight on me as I held her up and assisted her to walk gradually caused the bones in my feet to "give" and my hands to swell and other difficulties with shoulders, neck, wrists). Many times the patient and I fell.

3. Proper facilities. It's so very important to have the correct type of bed, easily moved chaises (wide, light-weight), wide mats for floor and yard. Really, I could at length suggest articles such as above and others I consider essential to the comfort of the patient AND to the nurse or attendant. We

did not in the home have the proper "helps" for the care of this patient: furniture and special wheelchair were needed, for example.

4. It is my firm opinion that TWO people are needed to care for a patient (in the home). I know I cannot again risk the injuries to myself attempting to care for such a patient ALONE. Great strength is needed. Great patience is a requirement. And a stable mental outlook is imperative to hold up under the unbelievable difficulties of humanely caring for an H.D. patient.

April 15, 1977

BETTY JUNE STEVENS
HESPERIA, CALIFORNIA

MARCH 20, 1977

My name is Betty June Stevens (Mrs. Jack). I am a 51-year-old Caucasian female housewife. I am not an "at risk" person in regard to Huntington's disease. I do wish to make my testimony known to the Commission.

My mother's family, Edward Grant Rathbun, and its members have endured years and years of extreme suffering from this disease. In my own lifetime, I have witnessed much of this suffering. I have observed the trials and tribulations of the families of two aunts, one uncle, six cousins and seven second cousins who have all been victims of the disease. Of these, only four are still living. Two of them are in the very advanced stages of the disease.

I have observed the frustrations of the families as they have tried in vain to obtain medical care and financial assistance. All, except one aunt, were finally placed in state mental hospitals for care.

In every case, there have been extreme psycho-social problems as the disease advanced. Many times the persons have tried to "end it all" by suicide.

In nearly every case, young families were left to be cared for by the remaining spouses or other relatives. Often, the families were separated to provide care for the children.

In every case, if the victim was the head of the family, it forced the spouse to seek employment to try to provide the necessary finances. This always presented more problems about caring for young children still in the family. It also presented problems about care for the victim, for often they could not be left unattended for their own safety.

Through the years, our family had felt this was an extremely rare disease. Very few doctors seemed to be aware of the disease. The doctors who were didn't seem to know where to refer patients for care.

In one particular circumstance, a cousin was brought from Nebraska to California seeking help. They went to a very well known and prominent medical clinic only to be told that she wasn't advanced far enough for them to offer any help. At that time, she was already to the stage where she had extreme difficulty walking and talking. She was already experiencing extreme emotional upsets. This seemed to be the common approach they received wherever they went.

I know the victims and their families need all the help that can possibly be offered. They need to realize that there are places and persons that want to help them and that they won't be "turned away." I feel it is of great importance that they be guided as to how to explain this disease to their children.

Definitely it has been proved that Huntington's disease spreads with each new generation. Therefore, I feel it most urgent that research be as intensive and comprehensive as soon as possible.

All of my experiences have been from a viewpoint of a relative but I would be very glad to offer my testimony or assistance in any way possible to help the research for a relief or cure of this disease.

Los Angeles, California

April 15, 1977

JAMIMAH SWEETEN
TURLOCK, CALIFORNIA

FEBRUARY 14, 1977

My husband died from Huntington's disease although it was misdiagnosed at the time because of a lack of knowledge on the part of the doctors. My husband died in 1970 after suffering over 15 years.

I have a son who is 45 years old this month who has just been forced into disability because of inability to function at his job--from this same inherited disease. I have two other sons, ages 36 and 30, plus a daughter who is 44. I have eight grandchildren who also face the gloomy prospect of inheriting this dreaded disease in their future.

I feel the greatest need is for research scientists to find the cause and a cure for Huntington's disease in order for our children's children to face the future with hope. This disease causes tremendous suffering on the families involved. Unlike cancer, this disease creates an unhealthy situation in the home as the emotions of children and spouses are involved as many times they do not understand what is happening to the parent and/or husband--this was the case in my home. For years we could not understand why my husband acted and/or reacted the way he did as we did not know he had inherited this disease from his father, who in turn had inherited it from his mother. A number of relatives are now suffering from this and I cannot help but beg you to put the scientists at work on this dreaded disease. I feel that the care for patients, needs of families, etc., are important but these I believe each family can handle themselves if only we can have the hope that our future generations can have hope in a cure or prevention of this disease.

I hereby urge this Commission to do all you can toward the cure and prevention of Huntington's disease--please don't spend money unnecessarily on care of patients, counseling, or therapy. We need a cure!

Thank you for hope.

Los Angeles, California

April 15, 1977

IRENE TALBOT
SAN MATEO, CALIFORNIA

MARCH 22, 1977

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it. I think this is a disgrace to think this terrible disease has been in existence over one hundred years and nothing has been done to eradicate it.

Los Angeles, California

April 15, 1977

ANN TAYLOR
TORRANCE, CALIFORNIA

MARCH 17, 1977

I grew up next door to a family during the 1940's. Met them in the 30's and in the 50's they moved across town. They continued to be like a second family to me. I would spend weekends there, etc. They were then what you would call upper class. There were four children in the Lechner family. I was Ruth's maid of honor when she married. Ruth is the third child born to the Lechner's.

I knew her mother was not well. She was acting kind of different, but no one seemed to actually say what the trouble was. Then the next thing I knew, Mrs. Lechner had been put in a home and Mr. Lechner had to sell his home to make ends meet. Mrs. Lechner was in the home 12 years before she died.

Mr. Lechner told Ruth on his deathbed what her mother died of. Since then, three out of the four children have it.

I can't believe how Ruth keeps her spirits up, but she does! She worked for years, and when she had to quit, she was something like 50 or 60 hours short of being able to collect Social Security.

There are drives for everything else. Why can't something be done financially for H.D. victims? Government paid rest homes for instance?

I probably haven't said anything here that helps, I know, but if the average person just knew what H.D. is, it would help!

GLENN M. TAYLOR
LOMITA, CALIFORNIA

FEBRUARY 21, 1977

My name is Glenn M. Taylor. I am 45 years of age, married, no children, and I am a school teacher. My mother has had Huntington's disease for about 20 years. She has lived longer than most. She has been bedfast for about six years. She cannot move, cannot speak--other than try to utter a word or two, has no control over body waste, and cannot ask for help when she needs it. Due to lack of speech, I am not sure how much of her mental faculties remain. I would estimate her weight to be about 50% of normal.

I think the most common cause of death with H.D. patients is pneumonia, caused by weakness and being bedfast. My mother was sent to the hospital twice with pneumonia. Each time her lungs were pumped out, it was cleared up, and she was returned. She receives good care in her convalescent home, but there is nothing that can be done for her or others like her.

I would like for my letter to be considered a plea for more money--or whatever it takes--for more research toward the cause and cure of Huntington's disease. Millions of dollars are spent on cancer research, cures for heart disease, etc. This is as it should be. But H.D. research is important too. H.D. does not kill as many people, but it's 100% terminal to those that get it. And to people like myself, because of heredity, we know our chances are exactly 50-50. I am consequently more concerned about H.D. than I am about cancer, or getting a heart attack.

About 1960 it was obvious that something was wrong with my mother. Her movements were affected, she did not walk straight, was nervous, had an uneven disposition, etc. I took her to a doctor in Huntsville, Texas. After a few days in the hospital he could find nothing wrong with her and suggested that she see a psychiatrist in Houston. I took her once, although even I knew it would do no good.

About 1962 I took her to an optometrist for glasses. He had her try to thread a needle. She couldn't, not because she could not see well enough, but because she could not hold the needle and thread correctly. He remarked that my mother was losing her finger dexterity. She was only about age 54 at the time, but he apparently did not think it unusual.

April 15, 1977

Not until about 1968 or 1969 did we get a correct diagnosis. My brother in the Air Force had military doctors in Las Vegas see Mother and they sent her to a neurologist near Riverside, California. He diagnosed her trouble as Huntington's chorea. He was very certain, and explained to us what to expect as time passed, and the inheritance problem for us.

When I entered Mother in the convalescent home here in Hermosa Beach, no one knew anything about her disease. The doctor that checks the patients, however, immediately read up on it and asked if I was aware of my own risk. I was grateful to him for that.

There are many problems for "at-risk" members of an H.D. family. I'll use myself as an example. I was a young adult when H.D. began affecting my mother. My dad was dead, and being the oldest child, I was expected to assume support. I did so for about fifteen years.

1. It is difficult to maintain a normal social life when a young man has a sick, dependent parent. Especially one with "abnormal" appearance and behavior.

2. It is almost impossible to build a normal life of your own--family, home, etc.

3. Society looks upon a young man caring for his mother (and living in the same residence) as dependent upon her.

4. Because of a dependent parent, unable to care for herself, I was not married until age 42. Even then (because of the "at-risk" situation) I was reluctant to do so. We have no children and plan to have none.

5. I did not put my mother in a convalescent home until I was afraid for her safety when she was left alone. Because of her fear of going to one it was the hardest decision I ever had to make.

I know the symptoms of Huntington's disease well. I know I am at the age when one looks for them. Whenever I drop something, or become angry or agitated easily, I worry. I have always tried not to let the risk bother me, but I know as I get into the age 40-55 years, it cannot be ignored.

Huntington's disease is a little known about disease, as far as the general public is concerned. Very few people know anything about it at all--and that includes a lot of medical people as well. Cancer, polio, heart problems, etc. are all well known about, but H.D. in its own way can be much worse. (1) It is terminal. The victim has no hope, while some cancer and many heart patients survive. (2) H.D. often drags on for 15-20 years. The victim becomes more and more helpless as time passes, until finally all body control is lost. Bowel and bladder movements are uncontrolled, and the victim can only lie in bed unable to move. (3) Since H.D. usually begins to show itself from age 40 to early 50's, many of the victim's most productive years are lost--and I am sure you can imagine the mental anguish that is suffered by both the victim and the family.

Research cannot be performed without money. Where the money comes from, other than from personal contributions I and other at-risk people make, is up to our Government, I guess. I would like to see H.D. publicized--television benefits like Jerry Lewis' Muscular Dystrophy, etc. perhaps. I do not have a lot of ideas, but I do know that there has to be research, and a lot of it, to find a cure for this terrible disease.

Thank you very much for reading my rather lengthy letter.

Los Angeles, California

April 15, 1977

JIMMIE C. TIMMONS II
FRESNO, CALIFORNIA

MARCH 21, 1977

I am 22 years old and my grandmother had H.D. My mother is at-risk and I think we need to have more research in the disease. With the help of the Federal funds and the knowledge of people interested in hereditary disease we should have a breakthrough before too many years have passed.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

BECKY TODD
FULLERTON, CALIFORNIA

FEBRUARY 27, 1977

I am a nurses' aide in a convalescent hospital, and I have a patient who has Huntington's chorea disease, and I don't know how to take care of her. This patient cannot be left alone when up in a wheelchair, as she thrashes around too much. She can't talk so I have no way to communicate with her. Can't the Government release a grant for research on this disease? We need to find out more about these patients so we can care for them accurately.

Los Angeles, California

April 15, 1977

DORIS J. UTIGER
WEST COVINA, CALIFORNIA

FEBRUARY 9, 1977

My letter concerns my father who was a victim of Huntington's Disease progressive deterioration. Initial symptoms were noted, but not understood, about twelve years prior to his death in 1975. However, until only two years before he died the family was unaware of his true illness. He had been mis-diagnosed as having Parkinson's Disease by two doctors in Chicago, and two doctors here in the Los Angeles area, and had been told nothing could be done for him. When finally a neurologist diagnosed Huntington's Disease and started the medication Haldol, which helped control the symptoms so well that he was still able to function fairly normally until six months before his death.

When we discovered Huntington's inherited quality, we were referred to City of Hope, Duarte, Ca., where we received very helpful genetic counseling, without any charge at all. We then found his mother's death in 1952 was due to the disease, but the family was never told, even though she had been diagnosed at the University of Iowa at Iowa City, Ia., almost five years previously! Consequently, I was 43 years old and already the mother of three teen age children. The genetic counseling information was given to them, but it was a difficult time for a 14, 18 and 19 year olds to find they may already be carrying the disease and to make decisions for their future life.

My mother was determined to keep him at home and care for him herself. This took an enormous toll on her health and strength. Fortunately, their finances were adequate so that she had help with the housework so she could devote her full time, night and day, to his care. The final months he was unable to walk or talk, bathe or dress himself, chew or swallow food, lost both bowel and bladder control, was emotionally unstable and confused most of the time. Since he was over 65, Medicare paid 80% of the doctor bills, but the medication expenses were very high. Every nursing and convalescent hospital in our area refused to even consider admitting him.

I consider myself fortunate that in my family, the symptoms of Huntington's Disease do not strike until after age 60, so at least a fairly full and normal life is possible until then. I also consider it fortunate that a coronary occlusion ended his life quickly at age 74, before the insidiously progressive symptoms of Huntington's Disease could kill him slowly.

Thank you for giving me this opportunity to speak on behalf of the control of Huntington's Disease.

Los Angeles, California

April 15, 1977

JAN VANDERFORD
PALM SPRINGS, CALIFORNIA

MARCH 8, 1977

Thank you for your time looking into this dreaded disease, Huntington's disease!

As an "at-risk" person, you must know many thoughts go through my mind.

My father is in advanced stages of the disease, and it is painful to watch the agony of the disease. I think it is becoming more than my mother can bear. I am concerned for them financially. At this stage of life, if Mom were to put him in a home, it would wipe out their life savings in two years. Dad is 62 and Mom only 60. The dreams they looked forward to are shattered. Of course, the worst thing is the agony, depression, humiliation and hopelessness that the doctors offer.

I am hoping that there can be more research into this and related diseases.

I am living now, with thanks for my health this very day, and appreciating it so much. My prayer is, of course, that my three sisters and myself can be spared from this horrible disease.

Los Angeles, California

April 15, 1977

FRANK J. VAN DONIS
NORTHRIDGE, CALIFORNIA

DECEMBER 30, 1976

In November of 1974 I was diagnosed as being an H.D. patient. My mother died with Huntington's disease. She was confined in a state mental institution at Alton, Illinois seven years before she passed away. My mother, Lillie, was one of seven children and I was one of five children. At present three of her five are afflicted, my brother being one of them. We are both unable to work and are on disability. Of us five children by my mother, Lillie, there are eight of her grandchildren at risk.

My wife's and my standard of living has been lowered due to my illness. I'm 56 years of age and it was never my intent to retire so early from my position as Service Manager of a well renown Cadillac auto agency. My health, mainly my nerves, made me realize something was wrong with my body and I would have to consult a medical doctor. But who and where do I start? August, 1972 an article was brought to my attention in the Los Angeles Times regarding information on Huntington's disease. We contacted Marjorie Guthrie in New York. One of her newsletters of May, 1974, told me where to go for medical help. This is how I met Dr. David Comings at the City of Hope.

In obtaining information, guidance and medical assistance, I have been very fortunate in my forethought in contacting Marjorie Guthrie. Without her accumulated information, I would have been completely at a loss as to my condition, and where I was going next. I feel what one woman has already accomplished has helped thousands find their way.

We, as H.D. patients, have a need for research, nursing homes, where patients can be watched and understood, genetic counselors, neurologists, nurses and complete information to doctors of the United States to further help to diagnose Huntington's disease patients and understand their illness.

Los Angeles, California

April 15, 1977

FINLEY VAUGHAN
FRESNO, CALIFORNIA

MARCH 22, 1977

I am a friend of a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

GLORIA WALTERS
FRESNO, CALIFORNIA

MARCH 22, 1977

I am friends with a family that has Huntington's disease.

In view of the enormous suffering Huntington's disease can inflict on individual victims, their families, and future generations, I urge your Commission to make every effort to combat and cure it.

Los Angeles, California

April 15, 1977

MRS. CHARLES WARD
SACRAMENTO, CALIFORNIA

MARCH 13, 1977

I am the wife of an H.D. victim, and to me, I don't think there is any other disease that causes so much suffering on the part of the victims and their families.

One of our greatest needs is a place where they can be taken care of properly--hospitals will not accept them and most of your better nursing homes will not accept them. If you are fortunate enough to get them in a convalescent hospital, it is just menial care and mostly the patients are restrained in bed 24 hours a day, so they lose what capabilities they do have.

Many doctors know nothing of the disease--so therefore are forced to put them in out-of-the-way places.

So I sincerely urge your Commission to make every effort to combat and cure this disease.

Los Angeles, California

April 15, 1977

FERN L. WELLER
PAUL W. WELLER
SIMI VALLEY, CALIFORNIA

MARCH 19, 1977

We were informed by a very dear friend that a commission for the control of Huntington's disease will be meeting in April. Our love and concern for Ruthie and her family and their future mandated that we write this letter.

It was a shock for all of us to first learn of this disease, especially since it affected a family, the Kruegers, that my family had been close to for generations. As a little girl, I remember seeing Ruthie's mother and knowing that she was very ill, but with what was always a mystery. Ruthie was kept in ignorance of the cause behind her mother's condition until told by her father in the mid-1960's. By then, she was married and had two daughters of her own, and since there are no "sure-fire" diagnostic tests to detect Huntington's disease until the symptoms of the disease appear (usually not until a person is in the late 30's, early 40's), there was no way for Ruthie to know if the disease had ended with her mother's death, or if she would later "inherit" it and pass it on to her own children. Her older sister was already showing symptoms of the disease, and not too many years later, Ruthie herself began showing outward signs.

At this time, it was also known that her younger brother had inherited it also; thus three out of four children were victims of H.D., all of whom had children. Whether these children have also inherited the disease won't be known until they, too, reach 30-40 years of age. Fortunately, the majority of these children have received the knowledge prior to having children of their own, but what a burden to have to carry. If they remain childless, the disease will end there with them for their family (H.D. doesn't skip generations). But what if they reach the age of 40 and still no symptoms appear, indicating that they haven't inherited the disease--the painful decision to remain childless to stop this disease would have been for nothing. After all, there is a 50-50 chance that a victim of Huntington's disease will pass on the disease to an issue, and there are many who are more than willing to take that chance. And who are we, who have healthy children, to censure them for taking this risk and knowing the joy, pain, pride and all the

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other emotions a parent feels in watching his/her child grow and being a part of that child's life? Would anyone deny them this too, when they've already been denied a normal middle age due to Huntington's disease?

I must admit that Ruthie accepted her inheritance with a great courage and continued her cheerful outlook on life--with one exception. She made it her cause not to turn her back on H.D. and pretend it didn't exist, but to inform as many people as possible--family, friends, neighbors--regarding the known facts of Huntington's disease. This is a very important part of fighting H.D. effectively--PUBLIC KNOWLEDGE AND AWARENESS. Because Ruthie wasn't afraid to face H.D. and not embarrassed to discuss its many related problems, she made us at ease, helped us to understand and accept her illness and never gave anyone a chance to pity her--there was too much admiration for her! It must be made possible for every person to have the opportunity to be informed and aware of the facts of Huntington's disease.

In our various social contacts, we have learned that only one other family already knew about H.D., and only because a member of their family was a victim. Ignorance of H.D. is rampant and a very real threat in itself.

Since Huntington's disease affects the motor nerves, often a victim's symptoms (slurred speech, unsteady walk, jerking or rocking motion) will appear to the uninformed as those of an inebriated person. With just a few of these symptoms as mentioned it should be obviously apparent that accidents are more likely to happen in the home due to not having full control over muscles. Communication with friends and family dwindles down to nothing as it becomes more difficult for an H.D. victim to hold and control a pen. People who do not understand what is happening to a victim feel uncomfortable being around them, thus some H.D. victims may feel like outcasts from society. (Again, a problem that can be corrected by public knowledge.)

Ken and Ruthie had to sell their home as it was just too much for Ruthie to handle herself and there was enough expense in treatment (what little there is) without adding to it the salary of a housekeeper. So they moved into a mobile home. Naturally, there is always the worry among husband, daughters and friends that something may happen while Ruthie is by herself.

Care in the home, even just companion type, is far too expensive today, especially on one income. Obviously, an H.D. victim isn't able to work once the symptoms become predominant, yet Ruthie isn't able to draw any social security (even though she worked prior to her illness and paid into social security), nor any other type of assistance or aid. With medical costs the way they are today and continuing to climb ever higher, what will happen when Ruthie requires more care than can be given at home by her husband and daughters? Where will she go? How can Ken possibly afford the expense of a private rest home, which at this time is their only alternative? Will the rest of the family have to help out with the financial burden as well as the burden of possibly inheriting the disease themselves in a few more years? These questions haunt an H.D. victim and drive some toward alcoholism, some in attempts to take their lives, and in other directions that provide "escapism."

It's a shame and a blot on society that there are so many on the welfare rolls that can work, provide and help themselves, but DON'T, when H.D. victims receive absolutely no Government aid and would give anything to provide it themselves, but are unable. Thus, there is a great need for some type of assistance for these people, in particular in the area of Government funded rest homes. After suffering themselves and knowing their family and friends suffer with them, can't the Government relieve them of one burden--to know they have a place to live when they reach the stage where their families can no longer care for them themselves, where they will be at peace, feel secure and well cared and provided for? We have come a long way from the days when H.D. victims were committed to insane asylums because the public was grossly ignorant of the facts surrounding Huntington's disease. That certainly couldn't be interpreted as "providing" for their needs. Can't we, as a Government, take another step towards enlightenment and set up pleasant rest home facilities?

It is our understanding that UCLA in Los Angeles, California, was conducting research on this disease, and until more is known and what is known is made available, how can doctors possibly give any type of medical aid to victims of Huntington's disease? The answers to this disease must be found and doctors kept up to date and informed regarding what is being discovered and learned so they may treat with the most up to date information.

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The four victims of Huntington's disease we've known are fighters, but we must give them something to fight with. It is so very difficult for all those involved to slowly watch their mental faculties deteriorate until they reach the stage of a vegetable or an empty outer shell, when memories come to mind of them when they were totally alive, vital people. During the last stage, our neighbor's brother didn't recognize, know or was even aware of anyone--as if he were in a catatonic state.

As of this date, I am 29 years old, married, a homemaker and former stenographer. My husband is 31 years old, District Field Service Supervisor for Dictaphone Corporation in Los Angeles, and out of gratitude and love for our three healthy children, we hope that this letter has been of help to the Commission. I have tried to give as complete a picture of the problems regarding Huntington's disease as possible in a letter, without going into too much detail to prevent loss of interest on the part of the reader. If we can possibly be of any further assistance to this Commission, please feel free to contact us.

Los Angeles, California

April 15, 1977

TWILA F. WESTERMANN
DIAMOND BEACH, CALIFORNIA

APRIL 12, 1977

It is with a great deal of renewed hope that I write this letter to you. Your efforts to secure information about the influence of Huntington's disease is greatly appreciated. The effect of this disease upon my family has been devastating. I cannot say that I would not have married my husband had I known about the familial disease, which was misdiagnosed as Parkinson's disease in my mother-in-law. After about a year, the diagnosis was made concerning my husband's two older brothers. (Since then, the other two brothers show varying degrees of severity of the disease.)

We were lucky to have known about it before we had our first two children, which we adopted. The adoption agencies were very reluctant about our fitness due to our family history.

Slowly I became aware of the symptoms manifesting themselves in my husband. I suppose one of the most difficult times is when part of myself wants to deny the possibility of the disease being a reality and the other part having to convince our associates that my husband's disinterest, his alcoholic-type swagger, and his inability to reason as well is not willful, but symptoms caused by the disease. The doctors were among those who needed my convincing. "Probably just emotional" was a phrase I heard more than once.

Finally, the doctor did take steps to seek a neurologist's treatment. Although this was a beginning, no effort was made to tell me about their findings or suspicions. I had to call them and ask. With my husband telling me that all the tests were negative, which in reality they are until the advanced stages, he must be okay. In desperation I made an appointment to see him. The doctor admitted he felt Ervin would need to accept the diagnosis on his own terms and therefore did not tell him of his suspicions. He was afraid of suicide if my husband would be told rather than him coming to the conclusion on his own. I expressed my fears concerning this arrangement, as I felt the doctor should be the one to tell him; and I, as his wife, should be the one to pick up the pieces, so to speak. At first he agreed,

then gradually the doctor said that I would probably have to tell him when the situation presented itself. I felt that was asking an awfully lot.

As the months went by, my husband constantly felt I was conspiring against him, as he tried to spend our insurance money on a florist business so he could support us, when, in reality, he would receive benefits continuously. Finally, in desperation, I gave in. Now we own a business which is a financial liability, which we would have a difficult time reselling because of its rundown condition due to my husband's mismanagement and my own inexperience.

Unfortunately my husband discovered a letter I had written to a close friend, to which I had poured out my heart. He became belligerent. He blamed me for "diagnosing" him. Instead of my being able to support him through this time I became the outsider and despised as well. If only the doctor had been honest with him. Luckily we have surmounted that hurdle and he no longer blames me.

Although I am thankful that our two adopted children were fairly easy to come by at that time, present-day families must face even the greater turmoil that even this avenue for fulfilling their parental instincts is not open to them. After years of worrying that the precautions we were taking against conception would somehow not be effective, I became unable to take the oral contraceptive because of nausea; other methods were not effective, as we now have a 3-month-old baby, which we felt deserved the 50-50 chance to a normal life, rather than eliminating it, as the law has deemed legal. Now that he does know, we can proceed with a tubal ligation, which would have been impossible to do before he was aware of his condition.

The only support I feel has been given me has been a handful of people who can only give reassurance that the frustration we feel is "normal." From others, worthless sympathy is standard procedure. Through CCHD this handful of people cares and can share a limited amount of support. They, too, have their own family problems which burden them. I hope this changes as it becomes better organized and can afford to provide proper counseling for its members.

During these last years of "existence" I have toyed with many ideas of how to escape this situation. Finding this not feasible,

I have turned to how to cope with these problems. At this point I am wondering if my original plan to keep my husband at home as long as feasible is a realistic venture. I'm wondering whether our children will suffer more if we remove their father from their daily lives or by having them be constantly reminded of how different our family is. Our daughter already hates to have her daddy touch her or eat at the same table.

Our son is developing "aches and pains" which he hopes will relieve him of household chores and give him extra privileges as his daddy receives. He naturally learns from his father that throwing things and hitting objects is a manner to express anger and frustration. A hole kicked in our living room wall by our son is an example. If a convalescent home is the answer or some live-in help, where does this extra income come from? Luckily my husband saw fit to take out an income protecting plan policy, so we have adequate income, but what do others do?

Such problems as when does he stop driving? When he takes a life or leaves himself or someone else disabled for life? Do we move closer to family, or is it less burdensome on a mother who has heart problems to have us farther away and not a constant reminder? Shall I build up the floral business and put the family to work in it so my husband can "keep busy" in whatever capacity he can for as long as he can, or shall I sell for a base price and travel while he can? Yet what does he do with his time between trips? All these questions and others are greeted by a sympathetic manner, but "I wish I could help you" is all that can be given.

I pray that your efforts for our cause will be amply rewarded. Thank you for the countless hours you have spent on this program on our behalf. I hope for you and your families the very best of health and happiness.

On the other side, my husband did not believe in wives working so I had not worked since our marriage. I was very lucky to get a job (with no experience) at Penney's; of course, working only part time. When my husband could no longer work at all we had to turn to county aid as my income was very small. Then one bill collector attached my wages and I was bringing home \$19.00 per week to feed my husband and two children. We filed bankruptcy and before we went to court our furniture was taken. After this and with the help of a nice relief worker at Wadsworth VA Hospital, we finally started to receive Social Security and a VA pension and straightened ourselves out financially to a degree. However, even now I could not solely support myself on the wages I make as crossing guard, which are higher than clerking in a store.

Of course, after the initial shock of being told about this disease we notified all my husband's family. And as my husband grew worse even some of his family have ignored him, preferring not to think about it.

My daughter is married and has three children and visits my husband quite often. It is very hard for her and although she tries to hide it, most certainly is a worry and heartache to think about. My son has many "hang-ups" because of this and the slightest attack of nerves and he is sure he has H.D. He has not married as yet and says he will never have children. He is also quite afraid to get too involved with one girl. It most certainly does not make a very happy life, but nothing we can say can keep him from worrying about it. Also, I understand the counseling is not that great. What can you say really? "Don't have children" seems to be top of the list.

Believe me, control of this dread disease can't get here too fast. These people become completely helpless but they are not idiots and being treated as such really makes me mad.

If you have read this, I sincerely thank you, and believe me, this is just a brief on the heartache and pain not only to the afflicted but the family.

Los Angeles, California

April 15, 1977

ELSIE WILLIAMS
BELMONT, CALIFORNIA

MARCH 1, 1977

Our family has suffered the ravages of Huntington's disease for many generations, and right now we can see symptoms appearing in a few of our younger members, which is frightening.

Because H.D. is so devastating emotionally, financially, and in all other ways to our afflicted family, our family urges this Commission to make every effort to find ways of combating it and of curing this disease.

It is really too painful for me to describe the pain and the suffering that H.D. has caused our whole family, but to give you a small insight I can say that those who suffered from it became totally unable to care for their families. As a result, children of these families had to be placed in foster homes while the parent languished in public institutions, suffering not only the loss of their family, but the loss of health and their ability to make a living.

Los Angeles, California

April 15, 1977

MRS. JAMES WILLIS
OXNARD, CALIFORNIA

JANUARY 14, 1977

It is very difficult in one letter to explain all the heartache and anguish caused by Huntington's disease. My husband was always very active and never too proud to accept any type of work to support his family without aid of any kind. At one time he worked two jobs full time.

When H.D. first affects a person he is usually thought to be drunk and people stare and children make fun of him. When my husband could no longer work nor drive a car, he rode a three-wheel bicycle and took food to a woman who was mentally ill and living alone. The food being purchased with money he could pick up by cleaning someone's yard or doing a few odd jobs. At this time he was continually hassled by the police and thrown in jail a few times with the threat of being hassled anytime they saw him walking on the street. I had explained the situation but was told I should keep him tied in the yard. True, the personality changes but you do not become minus all your faculties. Where once you were very modest, you will possibly become the opposite. You lose patience more with young children, although my husband never became violent. Also you find your "friends" no longer want you around because you might spill something or accidentally break something. And those you took loaves of bread to when they were having a hard time turn their backs and cannot be bothered.

When my husband could no longer handle a cigarette and stopped smoking--at that time seeing a neurologist at a Veterans Hospital--when he informed the doctor the doctor said, "You're a good boy. Here's a lollipop." Believe me, my husband knew what was going on. There is no need to humiliate anyone. Another doctor (when attempting to get him hospitalized) read his file, then asked me why the involuntary movement and what caused it. In my husband's case, he began to fall more and more frequently until he could no longer stand on his feet and only then did I try to get him into a VA hospital. I was told there was a list waiting and I should go to the county for help. After waiting two hours, I was told it would cost me \$300 to put him in a nursing home. My son-in-law called the hospital administrator at the VA and told him what was what and he was admitted in a few days. He has been there two years now, completely helpless and blind.

CAROL WITTOUCK
LAKEWOOD, CALIFORNIA

FEBRUARY 19, 1977

Almost ten years ago, I married a man whose father was seriously ill with H.D. With the naive faith of youth, I believed at the time that this could not happen to my husband. However, when we began to plan a family, we wondered, should we risk H.D.? We ruled out adoption, partly for emotional reasons, partly out of fear that a comprehensive physical exam would uncover some trace of the disease that neither of us yet recognized. We decided to trust to luck and I became pregnant with my first child.

It was about this time we felt we should inform ourselves about the probabilities, onset and progression of the disease. Very little had been written on the disease, and what material was available was contradictory. We were lucky?! Bob's Dad had been ill for a long time and was under the care of an H.D. specialist at UCLA Medical Center, Dr. Charles Markham. He explained the risks and the 50-50 chance of Bob's having the disease--and that should he have it, each of our children would have the same 50-50 chance. But aside from these facts, essential as they may be, what can genetic counseling for H.D. currently accomplish? True, it forces people--often young, hopeful, inexperienced couples--to face the hard realities of living with H.D. or the threat of it. But genetic counseling cannot yet determine which of the at-risk population can safely have children and which will pass the disease on to future generations because they themselves are victims. And outside of big cities like Los Angeles, which has few enough doctors trained in the problems of H.D.--very few, I imagine have genetic counseling at their disposal, or perhaps the money to pay for it.

At any rate, immediately after the birth of my first child and the death of Bob's father from H.D., I began to suspect my husband of showing early symptoms of the disease--increased twitching. Soon after these suspicions arose, Bob's 15-year-old brother was finally diagnosed as an H.D. victim. Couple these events with my emotional nature and I really had a problem on my hands. I felt overwhelmed and so very alone. I couldn't share my fears with friends or Bob's family as they might accidentally intimate their concern to Bob who, I felt,

April 15, 1977

couldn't handle such information, as he had always tried to escape from any mention of the disease. For this reason I could not even have my fears confirmed or dissolved by a qualified H.D. doctor. I wondered, "Why me?" and how I would manage, and worried that my children would, like Bob's brother, develop the disease as children before there was any real chance to find a cure. And so I hid in fear, anger, and guilt but always smiling to the world.

While I no longer live in dread of the disease, should it come--and it's likely that it has not as yet come--there has been a definite drain on our family relationships, particularly in my fear for my children's health and my resulting overprotectiveness of my oldest child. In the marriage relationship, anger and mistrust became a wedge between us as a result of my feeling the weight of this responsibility on my shoulders. Socially, I withdrew and am currently in therapy to correct these unhealthy symptoms. I don't claim that all this is the result of H.D.--since certainly I must have had the seeds of these imbalances within my personality--but the threat of H.D. did pose a unique problem and aggravated smaller ones.

Therapy is a costly program and Bob and I pay for 50 percent of it from our own income. We are lucky to be able to afford this luxury. Another financial burden associated with being at risk in this disease is the increased amount of life insurance we have decided to purchase.

Every family who has as one of its members a person sick with the disease was at one time at risk like we are. These are some of the problems that we faced and that others at risk might also face--but how much more complex they become for those who actually have to cope with the disease itself. I know, I've seen it.

We are all hoping you will help H.D. with your support and your dollars.

MAE Y. WORLEY
SANTA ROSA, CALIFORNIA

FEBRUARY 28, 1977

My sister, who is fifty-three years old, is in the last stages of Huntington's disease. I thank God her husband was able to keep her home until six months ago, when she could no longer walk or talk coherently. She is now bedridden, literally tied to a bed. Her movements are constant; it seems as though she is attempting to fight off or get away from this horrendous disease.

I am 51, an "at-risk" patient. H.D. is a 50% chance disease in more ways than one. I spend 50% of my thinking just trying to analyze my words and actions. Am I normal for my age? Do I think rationally? Am I unduly antagonistic and impatient with my family? Am I depressed because I worry about H.D. or is it H.D. causing my depression?

My father died of H.D. at age 60; he was mistakenly treated for "Addison's Disease" the last eight years of his life. He never had the choreic movements of H.D. He was very still the last two years of his life. He suffered from aphasia and was very calm except for an occasional fit of anger. I have often wondered if the large doses of per cortin and salt that he was given for Addison's disease may have in some way retarded the choreic movements.

My mother and I were quite sure my sister had H.D. when I read an article about the disease by Marjorie Guthrie. Then when it was finally diagnosed I went to see a leading neurologist in our community. He extended his sympathy but said that in his 15 years of practice, he had never seen a patient with the disease. I suggested it might have been a case of H.D. not being properly diagnosed in some of the patients. He definitely agreed.

I am willing to do anything that will help in the research and control of H.D., there are so many of us involved. My sister has four wonderful children and three grandchildren. I have five children and two grandchildren, and my brother has two children.

Los Angeles, California

April 15, 1977

Needless to say, the discovery of H.D. has had a terrific impact on our entire family. To see the change in my beautiful intelligent sister in just ten years is almost unbelievable. God help us all.

Los Angeles, California

April 15, 1977

MRS. LARRY YEE
SAN FRANCISCO, CALIFORNIA

MARCH 17, 1977

Due to the enormous suffering of Huntington's disease and its inflict on individual victims, their families and future generations, I urge your Commission to make every effort to combat and cure it.

I urge you to obtain Federal funds for the support and research of finding or diagnosis of at-risk persons and care for patients and families and all connecting with the Huntington's disease and its control.

Los Angeles, California

April 15, 1977

JUANITA ZIMMERMAN
LANCASTER, CALIFORNIA

MARCH 22, 1977

I have been a close personal friend of a family whose lives have been affected by Huntington's Disease. Having known this family for 30 or more years (since our high school days) I have seen the mother live and die with Huntington's and I have witnessed three of the four children develop this disease after being married and bringing their own children into this world.

I strongly feel more research into the causes, effects, prevention, and public education of this disease and tests to determine earlier in life if a person has H.D., before having children, are most urgently needed.

To see three educated vitally alive human beings reduced far below their capabilities is heart breaking. They have each gone through periods of family recriminations, misunderstandings, marital problems, psychological misgivings, and physical limitations due to this disease and I am sure a certain amount of outside embarrassment from people who do not understand their problem.

One was just one quarter or so short of being able to collect Social Security and now is unable to work or collect her Social Security. There is a definite need in situations like this for provisions to be made as the financial worry can be overwhelming. Also some kind of home where medicine, nursing care, and daily living could be accomplished for those who no longer can depend on themselves.

When I am with my friends I am amazed at the wonderful attitude they maintain even with all their handicaps, such as: coordination, speech, palsy, etc. All help given these victims of Huntington's Disease will be much appreciated.

I am a female, age 47, divorced, and a secretary.

Los Angeles, California

April 11, 1977

ANONYMOUS
SAN FRANCISCO, CALIFORNIA

NOVEMBER 10, 1976

Having worked in the medical field for twenty odd years, in Australia and here, I feel that Huntington's chorea is the most debilitating disease I have ever seen. Being powerless to help nearly kills me.

My husband died of it, also three other members of his family, and now my daughter has had it since 1964. Her husband divorced her, so now she is living with me. It makes me heartsick to watch the deterioration and nothing can be done to help her. The only medication that agrees is valium and that puts her to sleep. In the last few months she has been unable to communicate, masticate her food and even walk. In other words, she is a very sick person.

Unfortunately the doctors are unable to suggest anything to help these people. Why is it taking so long to come up with a drug that would relieve muscle tension? I realize the genetics have to be worked on, but please do find some medication first.

Another thing I want to mention, is there someone in a position to prevent the stuffy FDA from stopping a drug from coming into the country which would give these people relief?

No one should be able to block medical breakthroughs made abroad. Life and health is too precious to be played with.

April 15, 1977

ANONYMOUS

LOS ANGELES, CALIFORNIA

DECEMBER 7, 1976

Our family was not like other families; and I was preoccupied with wondering why this was so as far back as I can remember. Once I was reported to have taken my mother to task for not carrying me around "like other mothers;" but this is now beyond my recollection as I look back fifty years, in order to help the Commission realize what it was like for me to be at risk for H.D. and living with an H.D. parent.

I guess I have always handled my problem existentially, trying to get meaning, and somehow make valuable for me all my traumatic experiences, too numerous to recount. I was hurt and bewildered. Why was I unacceptable to so many people? Why was our family so isolated? It was rare that we had visitors; and especially if they were my visitors, why didn't they ever return? Why couldn't Mom and Dad come to school events to witness my accomplishments, which were not minimal to say the least? I learned to dread having my name announced publically because of the undercurrent of hostility I felt in the air. What I sensed one day became a dreadful reality when, what seemed to me like a whole audience of peers, teachers and parents, including my own, screamed at me in derision when I slipped, lost my balance and fell during a swimming competition held at the YWCA. I hit my head and this added to my shock. I was used to minor incidents like this, but I cried uncontrollably this time, drowning my sobs in the shower. No one came to console me and I was glad because I didn't want to face anyone. I had worked so hard to get my parents to come. I felt that it was necessary to pull myself together for the sake of my parents (10 years old). I must not let them know how much I was suffering.

I drove home with Mom and Dad in silence. We didn't talk about what happened. I refused to shed a tear in the presence of my parents. I had to force down my throat an ice cream cone that they bought for me. Later it was necessary to vomit it up when I was alone.

At age 44, as a graduate student studying for the profession of rehabilitation counselor, in a strange state, alone, and without family, I became quite certain that my mother's strange behavior, which had not become apparent to me until my early teens, had been attributable to H.D. A subsequent investigation which took several months later confirmed this hypothesis.

My mother's condition was not schizophrenia, a diagnosis I reluctantly made on my own in my late teens when her condition deteriorated to the point where I considered her dangerous to my physical safety. She had become violent and so I retreated from the scene, after trying fruitlessly to convince the family to seek psychiatric help. The reality of what was considered my problem was not even admitted; and needless to say, no remedy was suggested. I couldn't get along with my mother and it was even implied by the relatives that this was my fault. Events as I recounted them were believed to be false. Needless to say, my newfound acquaintance with the family secret hasn't drawn me to seeking any of them out. I have no wish whatsoever to contact them. I am trying to channel my justifiable anger and disappointment in people so that it will be a profitable thing for myself and others. Just as when I was a youngster I tried to cope through inner resources. No one can return to me what I was denied, some of the simple joys of life many people take for granted. No one can give me the close, tightly knitted family I have always dreamed about. At the same time, I consider that by paying a high price, one that I hope we will eliminate for all people in the future, I have gained certain valuable insights. I suspect that not all stories like mine will have a happy ending. God has been good. So far I haven't H.D. and I am trying to use my insights helping disabled people put things together a little better.

What were the circumstances in my family that needed to be put together? Most of the following factual information about my heritage was discovered by me six years ago. It was necessary then to reevaluate a lifetime of inexplicable and even downright tragic events with a new factual perspective, a perspective that, for whatever reason, had been purposely denied me.

My father who died at age eighty, clear of H.D., had always been at risk. His mother, sister, and brother all had the disease. I saw the brother. He had a very classical form of the disease with movements. I had never been encouraged to contact my father's family while he was alive. When I did contact them, I discovered that he had left home at age twenty, but had kept in contact through a post office box number. He apparently felt that it was necessary to keep his family history a secret. Dad suffered from seizures of the psychomotor variety at least from middle age onwards. Possibly these developed even earlier. When I witnessed his spells, I failed to recognize their true nature. I simply deemed it as inappropriate behavior. This certainly must have concerned him and must have contributed to negating the family image even further. It helps explain his reluctance to make

April 15, 1977

public appearances. It certainly compounded all of the family problems.

My mother inherited H.D. from her mother. There had been four children who reached adulthood and they have all been affected with H.D. Early in our family upheaval the disease had not become evident in anyone else. Mother was considerably older than all the others. My mother's aunts and cousins who were obviously "crazy" to me as a young child, had been introduced to me as her adopted family for which she had worked in her youth in the capacity of some kind of governess for the children. This was another deliberate deception.

The form H.D. takes, almost always, in my mother's family is rigid. This makes the illness less identifiable to an unsophisticated observer but not necessarily easier to handle.

I presently have two aunts with the disease. In our family there is longevity. My mother died at 76, having had the disease for thirty-six years, not the usual thing, happily. It looks as if her sisters will follow suit. One aunt has movements and this may explain why she would never see me and did not show up even for my father's funeral. I am afraid that I didn't understand or appreciate her motives at the time.

I conjecture that my mother got the disease around forty just after I was born. They had not planned to have a family and I was an accident. I had been Caesarian birth and there had been no complications. At age six a scoliosis of the spine was discovered in me and this necessitated surgery when I was nineteen years old. The deformity was arrested and I have had no serious difficulty, always having had to avoid heavy work.

Our immediate family was apparently the only branch that planned and limited their family. Our of thirteen cousins I now have, I know for certain four have developed H.D. and two others are beginning from all accounts to show symptoms.

I have never married and only for this reason did I avoid children. I suffered from social stigma for reasons unknown to myself most of my life. The situation was remedied when I left my home state for professional and educational reasons. I never had any insurance problems because, thanks to my parents, I could lie in good conscience when I was applying. I did, however, have all kinds of employment problems until I became professional out of my home area and out of state. I was a successful teacher for eighteen years and I now work for a state federal civil ser-

vice agency serving the disabled as a counselor in the vocational area. I did have trouble getting and keeping non-professional employment when I was a teenager in my home area. In retrospect, I came from an area where there are pockets of the disease; the disease is a part of the history of the area and feelings ran high forty years ago. I was looked upon as a menace. Knowingly or unknowingly, I could mess up the family heritage of another family (this is my explanation for what I experienced), something which, in its extreme, I hope is not typical.

Before I left home, I searched in medical books to try to diagnose the problem that no one would admit existed. My childish fantasy told me that I was living with a witch. Later I thought this stage of my development was significant in that this, in centuries past, was a conclusion that unsophisticated people believed and acted upon. I searched for answers in medical books. I was not completely satisfied that my mother was mentally ill. She did not always behave strangely. This was one of the difficult things about it all. I constantly had to readjust to the fact that there really was something wrong. Mother was a charming, friendly, socially-concerned and level-headed woman who never could remember chasing me with a carving knife, putting my hand through a plate glass window, hurling insults at me, throwing dishes about, and alienating my friends by misquoting me to them. I had serious doubts that I was dealing with schizophrenia.

In the medical library I found a small paragraph about a medical oddity called Huntington's chorea. The book had been hard to get for a young teenager and it was kept under lock and key. Some of what I read fit exactly with what I was experiencing. If mother had this, it would explain her behavior and the family problems. In fact, it would explain almost everything. I was gratified momentarily and a bit relieved that maybe my family did love me after all. Unfortunately, however, the account that I read in the medical library was incomplete (1944). It noted heredity but failed to mention the mode and late onset. Also, it did not explain that movements did not always exist. I didn't have the disease and mother did not have any movements. Also, there were no other affected family members that I knew of. I had been deceived in this but I didn't know this. Almost reluctantly, I had to abandon this new hypothesis which made everything fall into place. I had to conclude that mother was suffering from some functional mental illness and that out of some kind of fear the family could not face the fact. I had to go my way alone. I never tried to diagnose my mother to anyone outside the family nor did I ever feel an obligation to inform anyone of my mother's condition. In retrospect this caused some misunderstanding for me later on. Others assumed that I was doing what many Huntington's people do--hiding the facts. I think

Some positive relationships could have been established if I had really known how to be honest. I believe that my family and I suffered a lot of unnecessary anguish that might have been remedied if there had been available professional counseling, strictly confidential and within the special financial means of H.D. families. I regret deeply that I was not allowed to share the family sorrow. Family closeness and solidarity would have gone a long way to offset social alienation experienced in the larger community.

Based on my testimony, therefore, it may now be appropriate for me to summarize areas of concern I would like the Committee to report on. Much of these concerns stem, of course, from my individual experience with H.D. I see the Committee as valuable in being able, after weighing all testimony, to sufficiently generalize problem areas where Governmental concern and support may be of help.

1. There may be a significant but unknown number of people who have an H.D. problem; but because of deliberate deceit and/or ignorance, are unable to come forward, be counted and seek the kind of help they may need.

2. It would be foolish to assume that all H.D. risk children will have above average intelligence and will develop enough inner resources to deal with this catastrophe. Social deprivation with resultant psychological damage is a real possibility.

3. There very often exists, possibly related directly or indirectly to H.D., other physical and, of course, emotional problems in H.D. families.

4. In spite of what has been previously held, is it possible for someone to live and function with a mild but possibly unrecognized form of H.D.? This population could be considered feasible to benefit from specialized services such as vocational assistance which could help greatly in economic distress. (I will always question the fact that my dad may have had a mild case of H.D. If this were so, what would be the implications for me, even now?

5. There is certainly a need to educate the general public about the facts concerning H.D. Something unknown is always feared more.

6. It would be beneficial for those in the helping professions to be educated to realize:

- a. H.D. runs in families of every social class;
- b. Until affected an H.D. person can be functioning on a very high plain intellectually and morally;
- c. Victims of this disorder are not without individual worth and value to society. Even with their condition they are often capable of great insight, courage and social concern.

7. In order to safeguard individual choice and personal responsibility, it may not be desirable to control the disease by pushing for indiscriminate abortions and sterilizations nor to advocate universal birth control in the H.D. population. (In spite of all my traumatic experiences, I am glad to be alive and able to contribute my unique contribution to society.)

8. The emotional health and happiness of individual Americans will have an effect on the overall society in which we live.

9. It might be helpful if certain classes of disability (M.S., Pick's disease, H.D. and others) were declared catastrophic and special assistance given to affected families. At the present time, one has to have a good lobby going to influence legislation. Groups concerned with these illnesses sort of go it alone, and there are no groups for some people to go to.

Thank you for the time it has taken for you to read this testimony. Although I wish my name withheld from the public record, I am not reluctant to identify with the illness perhaps in a less formal, less public way. Feel free to ask me any questions about this testimony should you need to.

April 15, 1977

ANONYMOUS

SAN CLEMENTE, CALIFORNIA

FEBRUARY 7, 1977

In 1940, my mother was diagnosed as having "chronic, senile chorea, not-hereditary". Today, 37 years later, there are, including children, grandchildren, and great-grandchildren, 32 persons in my family who are "at risk".

Due to my mother having received an incorrect diagnosis (or was the doctor "sparing us"?), my sisters and brothers, the ones of us who still enjoy good health, are sitting on the edges of our chairs, praying --not only for ourselves, but most particularly for the sake of our children, and their children, that we have been spared.

Presently, I have 2 brothers who have diagnosed H. D., and 1 sister, undiagnosed and refusing to admit she also has H. D.

The older (61 years) of my 2 brothers, who was diagnosed as having H. D. 4 years ago, has tried 4 times to commit suicide, the last time by cutting his throat from ear-to-ear. Fortunately, this brother has no children. After nearly 2 years in and out of a neuropsychiatric hospital, he is presently living at home with round-the-clock nursing. Hopefully, his finances will last another year. Then he will undoubtedly have to go to a state hospital.

My other brother (57 years) is in a North Carolina Department of Corrections. A year ago, he was arrested and convicted of the felony of child-molestation and attempted rape. Shortly after he was arrested, Marjorie Guthrie requested Dr. Paul C. Bucy, Clinical Professor of Neurology and Neurological Surgery at Bowman Gray Medical School, Winston-Salem, N. C., to examine my brother. Dr. Bucy diagnosed this brother as suffering from moderately severe Huntington's Chorea. The sentencing judge recommended that this man remain in the prison system until such time as he is completely bed-ridden and it is necessary to place him in a health facility for care.

This brother spent 5 years in the Armed services during World War 2, and after receiving an honorable discharge, spent 20 years as an Executive in the Boy Scouts of America. The Veterans Administration states he is not "bad enough" to be a patient in a Veterans Hospital, and do not want to admit him, as he has a chronic, long-term disease.

Los Angeles, California

April 15, 1977

My sister, (60 years) whose chorea is more acute than my 2 brothers, refuses to admit she has H. D.

So, besides those 2 brothers and 1 sister, there are 4 more sisters and brothers, with 32 children between us, who are "at risk".

I do not wish our names to be used. I will identify myself as "H. G." I am a female, 48 years old, married, and a housewife.

April 15, 1977

ANONYMOUS
CALIFORNIA

FEBRUARY 8, 1977

For many years my husband and I have been close friends with a HD patient and his wife. We've stood by helplessly and watched this man go from a healthy, vital, energetic, talented wage earner to^a completely helpless, dependent being. Recently his condition started to deteriorate quite rapidly. He became so despondent that he finally tried to take his own life. The agony this act is causing is indescribable.

Since he was diagnosed as having HD he and his wife have contributed many hours and money in support of various HD chapter projects. He has been very cooperative in giving blood and tissue samples for research purposes.

We've known this man for over twenty years. Symptoms at that time generally went unnoticed. We did notice, however, that he had a rather irregular foot motion on the car accelerator. Out on the highway he would accelerate and then let up on the gas pedal several times within a 15 minute time span. We know now that this motion was involuntary. But at the time we thought he was just trying to be funny. Some time later he had debilitating headaches that were not at first diagnosed as HD related. He began having problems on the job that resulted in his going into another line of work. This didn't last very long. His symptoms were getting more pronounced and he was finally diagnosed as afflicted with HD.

Something must be done for HD patients and their families. Surely if we, as a nation, can put a man on the moon we can find the cause and cure of HD.

Los Angeles, California

April 15, 1977

ANONYMOUS
SAN DIEGO, CALIFORNIA

FEBRUARY 13, 1977

Huntington's chorea is one of the most devastating types of disease a patient can suffer. It passes through a family like a drink of water. To quote: "My husband died of Huntington's disease; his brother and their mother; now my daughter has it."

She has been a victim since 1964. Her husband divorced her, so now I am her conservator.

My daughter's condition is deteriorating. She is unable to speak, which is upsetting when the brain is still active. The rigidity of the muscles is terrible to watch and help. I must say very few doctors--if any--know what to do to help these people other than to suggest Valium.

To me it is annoying to think this disease, H.D., has been in existence over one hundred years and nothing has come to light to help these victims. WHY?

I would like to suggest something should be done; re special nursing homes for these people, not the type we hear and know about; re the treatment of patients; plus bleeding the family white for money they cannot afford. This question re nursing homes for H.D. patients is one of my greatest worries, because we are alone in this country. My daughter gets a small alimony from her ex-husband, and she would have whatever I can leave.

Having been in the medical field for 23 years in Australia and here, I feel very strongly that the Government without hesitation should release money for some good medication that would help these people to a more relaxed life; then work on the genetics. This terrible disease must be stamped out and soon.

Wishing each and everyone the best in this wonderful work.

April 15, 1977

ANONYMOUS

LOS ANGELES, CALIFORNIA

FEBRUARY 14, 1977

I am a 32-year-old, married, childless woman who is employed as a computer analyst and programmer in the aerospace industry. At the age of 13, I was made aware of H.D. (Huntington's disease) when my uncle was diagnosed as a victim. Since that time my father has also been diagnosed and is in a very debilitated state and nearing death. He has shown symptoms of the illness since the age of 10.

I try to attend all CCHD (Committee to Combat H.D.) and HDF (Hereditary Disease Foundation) meetings and am aware of the disease and its effects on victims and families.

My purpose in giving this testimony is to tell you of my family's experiences and those I have heard from other CCHD members. I have interjected observations, criticisms, social commentary, and, I am afraid, my own sense of frustration.

I have attempted to segment this letter into sections relating to specific or generic problems.

THE MEDICAL PROFESSION

I have found that doctors in general practice are not familiar with the illness. If a person should wish to discuss H.D., the doctor has to consult a reference book which often is outdated and contains misinformation concerning the hereditary aspects of the illness. Therefore, the doctor may give poor advice to at-risk patients concerning family planning, hereditary chances, and manifestation of the illness. (One doctor told me five years ago that only men can develop H.D.) Even if a doctor expresses an interest in the disease and is offered an HDF doctor's kit, he admits that he is too busy to avail himself of the knowledge.

It is very difficult to get good total medical care for an H.D. victim. The neurologists my family and others have encountered tend to be competent and understanding but are hesitant or unable to treat other medical problems (such as urinary, respiratory, heart, etc.). Therefore, it is necessary

to find a general practitioner or specialist to treat other medical problems, and one who is sympathetic and will not keep a restless H.D. victim uncomfortably waiting for too long a period.

Many doctors do not want to treat H.D. patients for other medical problems. They are afraid that if something unusual should happen that they would be sued. Some doctors even blame H.D. as the cause of all other problems.

It is also difficult to find a doctor who will make house calls. We once had to have my father admitted to a hospital through the emergency room service because he had a respiratory virus and was too weak to go to the doctor's office. The doctor would not come to the house to assess the seriousness of the situation, and the doctor felt it would be better to hospitalize him and see him there in case the virus was serious. So in addition to the doctor's hospital fee and the cost of hospitalization, we also had an emergency room fee to pay.

We had one very bad experience with a urologist who examined my father when he had a urinary retention problem (caused by the large dosage of tranquilizers). The doctor appeared to be very interested in the case; volunteered to talk to a neurologist in order to familiarize himself with H.D. and its side effects; and said that if my father ever needed his medical help not to hesitate to call him. The next time my father was in great distress with a urinary problem, the urologist refused to see him and told the nurse who was caring for my father that we should let the urinary retention problem progress into pulmonary edema and let him die. I realize that a heart attack, pneumonia, or some other illness may be a better way to die than to face the long, vegetative road of H.D., but I do not feel that other medical problems can be ignored, and I do not think that a doctor can make such a choice for us. Needless to say, the urologist caused a great deal of mental anguish for my father.

PROBLEMS OF LATE DIAGNOSIS

Due to the fact that there are no means of diagnosing an H.D. victim before there are definite symptoms (around age 40) makes it difficult to make decisions about having a family. When one has seen the disease and its manifestations in a parent,

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it is terribly difficult to consider a 50% probability adequate enough to guarantee that one will not become ill. Even if an at-risk person is optimistic enough to believe that through the advances of medical science his children may be spared or a cure or treatment may exist for them, one has to consider if he will have enough time to live a normal life and should he risk subjecting children to the problems of having an H.D. parent.

Another problem associated with the lack of knowing if one has the illness is the worry and psychosomatic aspects of the problem. One begins to think that every manifestation of irritability, lapse of memory, or involuntary muscle twitch is a sign of being a victim.

I wonder if a doctor could diagnose the illness at an early stage, if he would tell his patient the truth. Some doctors fear that an early diagnosis may lead to suicide, acute depression, or a great change in behavior. Some doctors I have met feel that in the case of slowly progressing illnesses it is better not to tell the patient so as to permit him to live a normal life as long as possible. Some of the at-risk people I know say that they do not want to know if they have the illness as long as no help exists anyway. Others would like to know so that they would not have children, enjoy life while they are healthy, and plan financially so as to provide for the prolonged medical care they will require later in life.

SOCIAL IMPACT

The social impact of the disease is enormous. Even though there are many sympathetic and helpful people, many people in our society lack sympathy, understanding, or tolerance for people who are ill.

My father suffered a great deal of verbal abuse from a few of his fellow employees. His boss and others who had worked with him for years were very understanding and gave him the easier work to do. Perhaps others were bitter about his being employed and permitted a limited work load. They lacked compassion.

One woman I have met, who has early disease symptoms, could no longer perform her particular type of work. When applying for disability benefits, she was told by a judge that she appeared healthy to him, and he felt that she must be able to do some type of work. The judge did not realize that due to

insurance company regulations concerning disability insurance and workmen's compensation, no employer is going to risk employing someone who is neurologically handicapped.

People tend to stare at H.D. people because they walk "funny," or squirm, or just do not appear to be normal. This quirk of human nature always has made my mother very uncomfortable in public. She did not like to go anywhere with my father anymore because it hurt her to see people staring at him. She no longer cared to entertain old friends. I have noticed that some H.D. families do begin to isolate themselves so as to "protect" the victim and to keep their problems private. With little provocation they begin to feel that nobody wants to be bothered with sick people, that nobody understands or cares, or that others should not be depressed by their problems.

Family members of a victim begin to make all the household and family decisions without consulting the victim. This is done in an attempt to spare the patient concern and worry which may aggravate his symptoms. The patient, however, begins to feel isolated, resents being treated in that manner, and sometimes feels that his family thinks he is incompetent. Needless to say, that the whole situation creates tension and stress for all family members.

Family problems become worse as the victim becomes less able to do things for himself. If only one person is living with the patient, that person has to sacrifice more and more of his freedom and time to care for the victim. This places the caretaker under more stress and provides very few outlets for that stress. The situation can cause illness, irritability, "paranoia," depression, or a sense of futility and abandonment in the caretaker. This causes anxiety and problems for all concerned family members: guilt, depression, a sense of helplessness, and an impact on their family life. Some cannot face the problem and avoid it by ignoring the victim. This causes more pain for the caretaker and concerned family members.

FINANCIAL IMPACT

Health insurance benefits are sometimes available to H.D. victims who have a spouse (or in the case of a minor, a parent) who is employed. However, not all group insurance plans pay for at-home nursing care. Fewer still pay for convalescent hospital care and most pay only a percentage of regular hospital costs. Also, most insurance plans have a maximum benefits clause

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which may be quickly depleted considering that H.D. is a long-term illness sometimes necessitating years of hospitalization. (One person I met had difficulty collecting her insurance benefits as they classified H.D. as a mental illness.)

In the case of a veteran, the VA in Los Angeles will provide some temporary care while performing diagnostic tests and research, but since H.D. is not a service related illness, they will only pay for six months convalescent care in a hospital of their choice.

Medicare benefits are available to people over 65 and may be available to others who qualify for disability retirement under the Social Security Act. In my father's case, he is ineligible because he is 55, and in the last ten years in which he worked he did not have four quarters of Social Security credit because he worked for an agency (Los Angeles County) which was exempt. This situation seems unfair to me as he has more than ten years credit in Social Security from prior employment.

In order to qualify for California's MediCal benefits, one cannot have over a couple of hundred dollars in assets and a home worth more than \$20,000. This would disqualify just about every homeowner in the Los Angeles area.

Therefore, it often becomes necessary to deplete one's life savings and property to qualify for any outside help. One woman who was receiving some sort of state or county aid was expected by the social worker to contribute her total salary to the care of her husband. When she inquired as to how she and her young daughter were to live, he said, "That is your problem."

The financial problems associated with any long-term illness can be the greatest burden on a family. It is very depressing for my mother who has to reduce her lower middle class standard of living still further. After having worked all of her life, she will reach age 65 with nothing left. It is especially frustrating when here in California we have so many people who have never worked and illegal aliens receiving welfare benefits paid for by us, and yet when we need help, we are ineligible because we own a commonplace house.

PHYSICAL LIMITATIONS

As the disease progresses, the victim becomes less and less coordinated. A swaggering walk develops. Finger coordination lessens. It becomes more and more difficult for the victim to perform simple tasks. In time, he has more difficulty walking, talking, swallowing, chewing, and controlling bowels and bladder. There is a great chance that he will choke or fall. In time he is totally bedridden.

These physical limitations present problems in caring for the victim. The person who cares for him must be physically able to support the weight of the victim while he is walking and be able to help him in and out of bed or up from a fall. If a wheelchair is used, it may be necessary to add ramps or enlarge doorways. Dietary considerations have to be made. Food must be of a certain type, easily digested, perhaps liquified or cut into small, chewable pieces.

Boredom is a problem. Reading is difficult for the victim as he may not be able to hold a book still or focus well. Most crafts and hobbies require more dexterity than he has. Frustration, depression, and self-pity can aggravate the symptoms. Television and radio, perhaps a drive in a car, become the only sources of recreation.

PERSONALITY CHANGES, MENTAL ILLNESS

I have not noticed any major personality changes in my father. Those that I have noticed are probably due to being ill rather than a direct result of H.D. Stubbornness and impatience were always traits of his and have become amplified since being ill. Mentally, he is perhaps not quite as sharp as he used to be (tranquilizers may contribute to that), but he does know and understand what is happening around him, and he remembers events from the past. He may become slightly disoriented for a short period of time when he is in unfamiliar surroundings.

It is interesting to note that when he was in the VA for six weeks some members of the medical staff assumed that he was no longer mentally responsible for himself. A psychologist who spoke at a CCHD meeting said that this is a common problem with doctors and nurses. She said that they do not realize that an ill person, who is mostly likely depressed at being hospitalized and away from his family, can become dis-

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oriented or uncommunicative; and they assume that senility and loss of mental faculties exist.

My uncle had a temporary loss of memory in the early stages of his illness and appeared to have delusions. This condition lasted off and on for only a few months and never seemed to occur again.

DIFFICULTIES IN OBTAINING ASSISTANCE

In 1958 my uncle was diagnosed as having H.D. Once my father learned that it was an hereditary illness, he lived in fear of also becoming ill. His only goal in life was to continue working so as to be able to provide a good education for my brother and myself. He felt that if he took good care of himself, ate properly, and did not strain or overtire himself that he would forestall any problems. Therefore, he would work hard at his job but would leave many household responsibilities to my mother. In time we began to notice symptoms of the illness but closed our eyes to them and told ourselves that we probably imagined there was a problem or his worrying caused psychosomatic symptoms. Once we had to accept the fact that there was a problem, our family doctor, who was one of the most dedicated and compassionate doctors I have ever met, consulted with a neurologist as to how to treat H.D. My mother and the doctor decided that it was best not to tell him that he definitely had H.D. but only a slight "nervous problem." In this way, he did not have to dwell on it, and I sincerely believe that he did not ever think (because he did not want to think) that he had H.D. until we told him when the illness had become more advanced and required other medical help.

Our doctor kept in touch with his neurologist friend and prescribed the drugs that were available at that time. When the doctor retired, the general practitioner who took over his practice showed an interest in the illness because one of his classmates was specializing in H.D. research in Montreal. However, as that doctor was rapidly expanding his practice to many offices and becoming less and less accessible, it became necessary to find another doctor. Living on the east side of Los Angeles has its drawbacks in that many of the more sophisticated specialists are located on the west side of town. Many second-rate doctors practice on the east side, and we and our friends have had a few bad experiences with some of them. No other H.D. family that we knew could recommend a doctor they had confidence in. So we turned to the UCLA Medical Center.

At UCLA the doctor prescribed new medication and my father appeared to stabilize for a while. However, upon using a new medication there often can be improvement or regression, problems in finding a proper drug dosage, and side effects. Yet when you question the effects of the medication or its relationship to other problems, none of the doctors seem to have any answers. We have been given conflicting opinions as to side effects and maximum allowable use. There does not appear to be enough correlation of data from doctor to doctor.

After a year of out-patient care at UCLA, my father became worse and my mother felt that it was necessary to have him hospitalized as she could no longer leave him alone. Having the impression that UCLA could not take him as a resident patient and not knowing anywhere else to go, she took him to the Veterans' Hospital in West Los Angeles. I have heard that there is a ward for H.D. victims there, however, administration sent my father to the neurological ward for evaluation.

The typical procedure at a VA Hospital (and we have been to two of them) is to keep out-patients and new patients waiting for hours before they are treated or evaluated. Someone being evaluated often has to go from one medical department to another which means going to many different buildings. All the waiting and walking is hard on an H.D. victim, not to mention other people with serious illnesses. Also, the staff was rotated on a monthly basis so it is likely that you will have more than one doctor in charge of your case. My father was placed in a ward of patients of differing neurological problems. The building was old, lacked window coverings, air-conditioning, and generally depressing. The staff ran a series of tests on him, prescribed higher doses of medication, and prescribed physical therapy. One test they wished to give required a consent form. When my mother hesitated to sign it, the doctor implied that they would release him immediately. The doctor admitted that the test was not required for diagnosis or treatment, but was to satisfy his curiosity and perhaps be useful for research.

The care at the VA was barely adequate. He was bathed but his soiled clothes were not changed often enough. He was confined to bed and no one helped him with his food. Other patients who were physically able to care for themselves assisted him. The VA stated that he could not stay there but they would pay for six months in a convalescent home. Since the list of the homes they have available to them did not contain one within

fifteen miles of our residence, my mother asked if she could have a week or two to look at the homes they suggested. The doctor who was head of the department (they also rotated department heads every six months) agreed to this, but the social worker assigned to the case and somebody in administration would call her frequently urging her to hurry and take him out of the VA. When she found out that her health insurance would pay 80% of at-home care and was able to find a nurse to care for him, she decided to take him home and informed the doctor that she would pick him up at 7:00 p.m. At one o'clock someone in administration called and stated that if she was not there on time, they would put him out on the curb. The doctor could not understand why anyone would have made such a call, yet it happened.

This is not necessarily an indictment against the VA, but at the time (and maybe they have changed) the constant rotation of personnel, the loss of files and lack of good administration, the lack of empathy, inexperience of doctors, and hours of waiting for out-patient care do not make it a desirable medical facility.

It was through a CCHD meeting that we learned that the City of Hope treats H.D. victims but does not have hospital care for them. However, when we called the City of Hope we were told that they did not treat H.D. people. So we had to get the name of the doctor from the other CCHD member and call the doctor directly. In that way, we were able to get good out-patient care from a doctor who is experienced in treating H.D., has sympathy and understanding for his patients and their families, and does not think of them as guinea pigs.

Problems exist in getting financial aid through governmental agencies. You hear from friends that someone in the same circumstances as yourself is receiving some form of aid or hospitalization. We call the agency and speak to someone who tells us the eligibility requirements and make an appointment for an interview. Once we have filled out the forms and been interviewed, we are told that we are not eligible for any benefits and whomever we spoke to gave us the wrong information. Meanwhile, we have wasted a day, lost income from work, and feel more frustrated and depressed.

Nursing home care is expensive. Anyone who has ever known anyone who has been in a convalescent home does not want to send an H.D. victim to such a place knowing that they will not get the individual care that they sometimes require. Hospitals

fear that an H.D. person is prone to falling and do not want them moving around even though some exercise is good for them. Convalescent hospitals employ teenagers and people with no training. Our state hospitals are not any better as the recent scandal concerning patient abuse and deaths can verify.

Most families want to keep the victim at home as long as possible, but this puts a tremendous burden of responsibility on the caretaker leading to their own physical and mental problems. I really feel that a hospital that specializes in the care of H.D. and similar type illnesses is required. There would be much less pressure and worry for H.D. families if they knew that such a place exists.

SUMMARY

Perhaps it appears that I expect too much. I realize that there are many sick people in our country who do not receive good medical care. I know that there is nothing that can be done about the lack of consideration in people and the poor attitudes that exist in the medical profession. I also realize that governmental agencies are hampered by bureaucracy and inept personnel. However, considering the hereditary aspects of the illness and the extreme disability it causes, I feel that extreme means are necessary to do something about it. I also feel that in the United States it should not be necessary to go bankrupt before financial help is available.

Perhaps people my age are the first to realize the full impact of Huntington's disease. In the early part of this century the disease was seldom correctly diagnosed and the hereditary aspects were not fully known. People did not live as long, were killed in wars, or the illness was thought to be the results of alcoholism or some other problem. It is only recently that it is diagnosed and that the hereditary chances are reported to be 50% for direct descendants be they male or female.

It is people my age who have the problems and heartache of having a sick parent, the dread of becoming a victim, and the burden to consider all this carefully before procreating.

In spite of the social, financial, and psychological impact of H.D. on the family, I feel it is necessary to live

Los Angeles, California

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in hope that the cause of the illness be discovered; that some treatment can be found, if not to cure then to at least relieve the symptoms; and that help be available in caring for those who have to be hospitalized.

ANONYMOUS
BURBANK, CALIFORNIA

FEBRUARY 8, 1977

CASE ONE

BEGAN:

Ever since Mr. S. (then age 40 years or so) got hit by an uninsured Chicano motorist he began to deteriorate rapidly in H.D. Doctors speculated the blow to the head may have helped bring it on quicker.

COMMITTED:

After a few years (to age 45) of living at home, he was committed to a state mental hospital (1955) and died there in 1963 (age 53).

PROBLEMS:

MEDICAL: Getting a diagnosis; was a "guinea pig" at a university for anything they wanted to try. He was willing to help find information for research; family doctor urging sterilization of both male children. In those days, it took opinions of three doctors before sterilization could be done. Nowadays it's much simpler, so boys were told to wait until 21 years old. No treatment apparently known by local general practitioner. When committed he was lucid and for years afterwards; this I surmise from subsequent conversations reported to me; and, however, care is not of more than a custodial nature, since staff is told he is "incurable, terminal and getting worse." No therapy is routinely used to help H.D. victims.

SOCIAL: Society - Any loss besides cost of custodial care? This man had his name on six patents. He was working on his doctorate when he was involved in the car accident. He was a chemical engineer, graduate of a university. He was the discoverer of a use for diatomaceous earth, now used as he discovered in filters and purifiers of all sorts, most commonly in pool filters.

To Himself - No direction or guidance available except for researchers giving him all sorts of tests for information for their research, but no counseling, no warning of how to cope as

deterioration continued. Loss of his home life when he was committed.

His Wife - They owned their home and a large apartment house outright when he died and had money in the bank. She was more fortunate than most who are impoverished by loss of the breadwinner. However, she was of average intellect, worked two months for the phone company once, and was a housewife. She relied solely on guidance of her general practitioner who was very pessimistic. After putting away her husband, she expiated guilt every other weekend driving up to the hospital from home until her husband's death. All holidays, his birthdays, and their wedding anniversary she takes flowers to the grave and cries. She will do this until she dies.

She is out of contact with relatives from her side of the family, mostly all dead now, and her brother-in-law (who was married to her husband's sister) avoids her as does his son. His eldest son: Age 41 now. Not married. Lives with mother. Had a nervous breakdown in college after completing 3-1/2 years. Never went back to complete for degree. Never took CBS job offer because it meant moving from home and leaving mother. Hysteria displayed instantly if H.D. or anything related to it or research or Commission hearing, etc. is brought up. Has an unstable work record (but employed now), not from lack of knowledge, but, I believe, because of the image he projects to people; a closed mind, teetotaler, picky over collecting small papers and labeling things.

How does he contact the outside world? Listens to shortwave broadcasts and mails letters to stations around the world for a "verification" that he heard them. I helped him start a "shortwave file" in the pamphlet section of the local library and he is very proud of keeping it up to date. Encouraged him to join a shortwave club. He finally attended a meeting, but his mother said she'd go with him and did.

His younger son-- Unmarried until age 29. Living life of a noncelibate bachelor. Active in airlines and travel field. Refused to even own any furniture until after third year of marriage. Reason: "In case I have to move...have to leave..." After six or seven years of marriage he began to think of finding a house and buying it. Engaged three times. Broken off in two cases by the woman with stated reason being not wishing to live with H.D. In one instance was engaged to an R.N. who had an H.D. male patient as one of the patients she cared for. It

was a long and serious engagement to him. The woman broke off and had just learned she was pregnant. My husband begged her not to go; she went to the next state for almost a year. When she came back there was no baby. She wouldn't communicate with him. He'd been calling on her mother all that time and waiting only to have his letters returned and never did learn exactly where she was.

It was several years after his marriage that he went past her parents' home without even unconsciously looking for her, or glancing at the house. That was about 15 years later.

Present Status: Married. Has a vasectomy. No children. How was vasectomy initiated? Urging of his mother (who paid for it) after several months of marriage and mother's urging and urging of two R.N.'s and one doctor his wife worked with who all kept urging her to sign for him to have it, and signature of his mother's general practitioner who never gave them an appointment, just mailed "okay" to surgeon.

He never said he really wanted a vasectomy, just kept asking his wife if it was what she wanted, when they were in the surgeon's office, and they did not talk about it much when at home.

Attitude Toward Vasectomy: Both husband and wife feel if family had counseling so mother would have been more rational and husband and wife would not have been subjected to negative public opinion (through her work) and ignorant God-playing general practitioners, anxious to keep favor with the old mother, then they feel they would have not spent so much time later in their marriage paying two different psychiatrists to find out what they wanted. The wife gave in because of traumas in her life that made her swear she'd never have kids to protect them. Pushing her father away when he was still holding her and watching him go through the door to shoot himself, then having a step-father who after years of extreme punishment and repression molested her and she was too afraid to fight back.

Vasectomies are "giving in" this couple feels. "Okay, H.D., you'll get me so I'll quit right now. No normalcy in my life. I'll cut my responsibilities so when I get sick, I'll be all ready to die."

To have kids, on the other hand, we've come to feel is a positive attitude. A "keep fighting" attitude. A chance to be

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good parents, and even if the father gets H.D., a chance to teach the kids "Love and will," as Rollo Ray says, "are the most important things."

But the lack of any knowledge of any organizations fighting H.D. or any research being done on H.D. at the time, of knowing of any one else with the problem, and reminder that "it's incurable," combined to influence us to have a vasectomy.

Memories: Social stigma due to lack of public knowledge:

1. Ridicule at school by kids who'd seen his father "drunk on the lawn."

2. Being avoided after trying to explain it was an hereditary disease.

3. Being embarrassed to take girls home when first dating and father was still at home.

4. Taking one girl home after school only to have her start laughing at "the drunk watering the lawn."

5. Father being hassled on walks by police for being "drunk."

6. Father smoking in his favorite chair and coming in to find chair in flames and father in it because he couldn't get out and hadn't been able to shake out the match from the cigarette.

7. After chair incident, remembering father being declared a danger to himself and those around him. (This he learned later.) He was taken out to the backyard one day by his mother with his older brother. His father was in the front yard watering the lawn as usual (one of the jobs he loved to do--to get outdoors and do something useful). There was the sound of a car and screaming from his father for help. His mother tried to hold both boys but while the older one stayed frozen there, the younger one ran out to the front yard to see two men in white coats trying to drag away his father. He helped his father fight them off until his mother came running out, and said, "Stop! I told them to." It was a complete surprise to the father as well as the boys. It's taken years of marriage to finally begin to convince the man not to kill himself at the

first diagnosis of H.D. if he gets it because his wife couldn't do that to him.

Attitudes of Doctors Encountered:

1. Wife's doctor--never heard of it. Finally found two lines in one of his textbooks.

2. Surgeon--sympathetic; most unwilling (never said so but now I feel he really was) to perform vasectomy but could not reach either husband or wife through their individual scars. Just said he'd do it.

3. Gynecologist--wife and husband went to twice who was most reluctant to sign vasectomy okay, but wouldn't say "yes" or "no" to respond when asked if he recommended not doing it. Was told by husband that if just one of the three doctors came out and said he didn't recommend it at all, he wouldn't have it done. No one expressed an opinion except:

4. Mother's general practitioner--who wouldn't see the couple, but mailed Rx for vasectomy to surgeon. Just recently (within last two-three years) when a woman asked me for any doctor in that area or city who was familiar with H.D. I gave her his name, remembering he treated Mr. S. for years. He told her he never had any H.D. patients and wouldn't take any.

5. Swedish doctor--never heard of it. When mentioned, claimed husband was making up the disease.

Attitudes of Husband and Wife Now: FIGHT. Get public support so people respond kinder without shunning patients. Husband has been area representative, officer, and trustee of CCHD. Wife has been committee chairman and trustee of CCHD. Both are still in all three local chapters of NHDA, CCHD, and H.D. Foundation. Both have helped patients, donated tissue to research programs, visited patients, mailed and delivered H.D. materials printed by organizations to various doctors and patients' families.

The ultimate way to fight H.D. is the reaffirmation of life and optimism, having children. It's not being defeated or depressed. Hope of a better inheritance.

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Ironies: All the money the surviving wife of Mr. S. now has was earned by her husband, but because she "doesn't want any of that literature" coming to her house for people to see, she has not contributed to any H.D. organization which might eventually help her sons or serve as a worthy memorial to her husband.

She contributes very conscientiously to the Arthritis Foundation, which she suffers from.

She had her husband committed because she felt she couldn't take care of him. They had enough regular income to afford a nurse. When the woman's mother needed care, she took her in and had her live with them.

Lack of knowledge, fear, no supportive companionship, shame because of public reaction, all contributed.

CASE TWO

Mrs. V.B., sister of Case One, got H.D. around late 30's. Died around late 50's or mid 50's (two to three years ago).

Husband moved to mountain resort, bought a cabin. Kept wife there until she died. He commuted down the mountain to work in Riverside. She had no social life, no known nurse care.

Husband still will not associate with former sister-in-law by marriage, each feeling that the other acted in a wrong manner in dealing with the H.D. spouse, but neither knew of any public agency offering any help or counseling or therapy for their problem, and both thought they were alone with the problem. No known organizations. The son of this woman very rarely visited.

CASE THREE

At-risk Son of Case Two. In mid-40's. No symptoms yet. High school coach-gym instructor. Never communicates with relatives, active in H.D. organizations, nor with ones who had H.D. in the family.

In second marriage. Did not invite "H.D. activists" to wedding. When they asked him why he said he didn't want them talking about H.D. They swore they'd have no reason to bring up the subject at a wedding.

So far as is known his second wife knows nothing of H.D. His kids know nothing of H.D. His first wife never knew about it until when she thought she might be expecting a third kid and she asked H.D. activist counsins' wife why they had no children and counsins' wife thought it was okay to answer and did. She said, "No kids because of H.D. risk." Turned out woman knew nothing of H.D., didn't know why they never saw her mother-in-law in the mountains, and had never known what exactly was wrong with her. They were divorced shortly after that.

Only communication with man and second wife is in replies to Christmas cards second wife mails out every year.

Will try to mail copy of Commission request for information.

CASE FOUR

Mrs. B. (fake initial). Woman in early fifties; one adopted son. Company dismissed her. Said she couldn't do the work. Didn't qualify for unemployment because not diagnosed yet, when dismissed for falling and dropping things on the job. When weakened by H.D. woman picked up a lot of other health problems. Expensive. Husband couldn't afford medical care. Couldn't afford days off work to fight for her disability work benefits. An interested insurance agent started the ball rolling. Case was appealed. Used medical testimony since obtained, R.N. in uniform pushing Mrs. B. in wheelchair into court (was local CCHD member borrowed for the occasion). Local area representative of CCHD came in suit with literature for court about disease and problems. Local state assemblyman called judge beforehand and talked about it (local CCHD area rep's wife had contacted him--he'd never heard of H.D. before). Woman's case was reversed and all back benefits paid her and her son as a dependent of hers. Finally got her on Meals on Wheels list. She lives at home, has Medicare now, etc. Only qualified for MediCal because of expenses due to other problems.

Follow-up of one of woman's neurologists. One young neurologist came into hospital once to see her, didn't give her any tests, didn't do anything but walk over to her bed, look at her and say, "You don't have H.D." She'd been diagnosed by several different men, specialists, for last few years as H.D. patient. She offered him one of the doctor's kits she had made up to pass out to interested doctors at the hospital (supplied by local

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CCMD representative) but he refused it. Said as she didn't have H.D. he didn't need the kit and no other patients of his had H.D. either. I couldn't conceive of anyone in the medical field being that closed minded to useful information. Hope he NEVER gets an H.D. patient.

CASE FIVE

Mrs. W. (fake initial) about 53 years old. One young married daughter, a dancer, who travels and can't care for mother; one 18-year-old son she was living with, who wouldn't speak to Mrs. W.'s guests when they called on her and who just ignored his mother or left the apartment or turned up the T.V. when she spoke to him to introduce guests, etc.; one daughter in own apartment attending junior college.

Mrs. W. really needed someone to fix her good meals which she was not receiving. Teenaged boy couldn't cope, didn't cook, and tried to disassociate himself as much as possible.

Kids had spoken to mother before about her living in a rest home, but she got very upset and wouldn't hear of it. Said they didn't "love her" and were just interested in getting her out of the way. She didn't hear anymore about it. Next she received an eviction notice on her apartment for "nonpayment of rent." She had always paid it, always given the money to her 18-year-old son to pay it. She called a friend, upset that she was being evicted, upset that none of her kids were helping her find another apartment, even though she kept calling them and was worried about "being put out on the street." Her friend worked full time and had a child to take care of, but promised Mrs. W. that if her kids wouldn't help, she would drive Mrs. W. around apartment hunting. Next time she called she couldn't get hold of Mrs. W. The friend finally made contact with one of Mrs. W.'s children, learned Mrs. W. was in the hospital for depression. Went to see Mrs. W. Mrs. W. said she was (not sure) told she was going to a rest home. Mrs. W. sobbed, carried on, said "Her kids didn't love, etc., etc., no one paid any attention ..." Was put in local hospital for depression. This gave kids opportunity to empty her apartment. When friend came to see Mrs. W. in dismal surroundings where she was to stay until cheered up, friend asked how they were helping Mrs. W., what was the treatment? Mrs. W. responded that they were just giving her her regular daily medicine that she always took anyway. "Anything else? Any therapy?" "No, nothing else."

Mrs. W. was released, sent to retirement home to live. Sobbed, carried on (figured this is the beginning of the end), called suicide prevention one night and said she was thinking of throwing herself in the pool, her kids didn't talk to her, love her, etc.

"Center for Prevention of Suicide" now had a record that she was a danger to herself. She was put in a state hospital. Upon visiting her (your name must be on the list or you will be told there's no record of such a patient there) she wailed that her 'Kids didn't love her, why I ever called Suicide Prevention and say I was going in the pool? I'll go anywhere, get me out," etc.

She'd always gotten her hair done regularly and dressed nicely before. She looked like a different person in a single shift with white roots almost 3/6" long and sticking in different directions.

Last time I visited her I had a hard time getting an attendant to answer the bell and unlock the ward so I could enter. When I did enter, I couldn't find her in any of the usual places; T.V. room or visiting room, and I'd called an hour beforehand to give them time to clean her up and prepare her for a visit. The aide located her, unlocked her bedroom, and helped her slide out of the high hospital bed she was in. By using the aide's help she could slide and reach a footstool the aide put by the bed so she could get out and come into the visiting room. With all the muscular coordination problems she's supposed to be developing, I was also surprised that she was in her stocking feet trying to walk on that shiny linoleum floor.

Mrs. W. was telling me that a minister came by to see her in the hospital and tell her she has an "incurable disease." That's correct and it was thoughtful of the staff to send a minister by, if they were responsible, but she doesn't need to hear that. She wants out and she has many productive years ahead of her before she reaches the stage most H.D. patients are put in mental hospitals. She also knows there's research going on all the time. She says, "To go back to the rest home would be heaven." But the rest home doesn't want her back now that they got her out. She says her kids say the rest home doesn't want her and "there's no where else they know of." This woman has her social security benefits and \$525.00 monthly coming in.

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Her eighteen-year-old son is now in his own apartment. He's a senior in high school. With an income like that, I'm surprised that her kids haven't been able to find a place where she could go. I don't know who handles the money.

Commonly, when a patient is in such good shape as Mrs. W., the relatives don't mention H.D. when getting the home to accept the patient. Only when it's a fact that they've moved in and it's difficult to turn them away, is the home told. If the patient can do as much as anyone else, to take care of themselves, then the home can see it and is more inclined to accept them.

CASE SIX

Mrs. S. (fake initial) has a daughter, now 15, who has "Neimann-Pick" disease, a problem similar to Huntington's Disease: muscular loss, brain deterioration, convulsions. The girl was 12 when she got it. It's hereditary but not as dominant as H.D.

Mrs. S. was in the office I work in as a patient and was even reluctant to tell me about her daughter's problem even though I am a health professional. She told me though because she wanted to see if we would take her daughter as a patient.

This woman almost burst out crying when I told her I belonged to three organizations interested in hereditary disease and its cures. She wasn't so stunned at learning they exist (the organizations) here in Southern California as at the fact that I told her they are comprised of families who meet together to work out the related problems and learn from each other and talk. Mrs. S. felt she was alone in the world. She is divorced. Her husband pays all the medical bills and for the nurse-attendant with her daughter.

Mrs. S. has the common enough problem of not having had any regular association with any relatives since her daughter began deteriorating. One brother just told her during this Christmas season that they found it too hard on themselves to look at her daughter and that it disturbed their own daughter and encouraged "the fear that it may happen to her." The odds against that being true are ridiculous--if I remember right 1:100,000--but this is the excuse.

SUGGESTIONS

1. Background investigations for various jobs (i.e., L.A. Police) should only be allowed to investigate the applicant, not check out parents, where they died, etc.

2. Every job with a pension fund tries to ditch employees for medical reasons. The Government office my friend works in dismissed one man because they said his varicose veins affected his "ability to do the job" because he stands up on part of the job. This man had never told anyone but the company medical insurance application form about his varicose veins. It should be against the law, "invasion of privacy" or something, to tell the employer, a third party, about confidential medical facts. If they decide you can't do the job it should be based on performance, not prejudicial information.

3. When one is dismissed for not being able to perform a job satisfactorily, one should be able to file for disability up to several years later without needing to put out a lot of money first.

Example: A woman was dismissed from her job, told she couldn't perform. She was falling and dropping things. After the time allowed for an appeal was up, I believe several years, she learned she had Huntington's disease. Committee to Combat H.D. for Southern California had a vice president who had a cousin who's a lawyer. He came out to see the woman for nothing. He reviewed the case, but after learning that she had a case, she learned to file an appeal would cost \$200 or \$300 which she didn't have.

4. There should be a provision in the Social Security laws for occasions like this. There are provisions for family group counseling in Ventura County for families breaking up over marital troubles. There should be therapy groups for families suffering traumas from catastrophic or hereditary illnesses. Groups with similar problems may be able to give each other suggestions as to how they dealt with various problems. Such people, from my findings, are reluctant to open up until they find a listener familiar with their problems and who can show them others with like problems, and that people are dealing successfully with them.

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At present the only counseling available is private. Knowledge of H.D. is general, not exacting, in the case of most private therapists. Groups could teach the therapist while the presence of the therapist would discourage "one-up-manship" stories as some may want to tell from time to time.

5. Once my husband and I told a life insurance agent about H.D. We wanted to get rid of him. We told the odds, etc., life span. He was horrified at almost having signed us up. He checked and told us just not to mention it when we applied. There are pools in the State of California for insurance companies to pick hard-to-insure motorists out of and force the companies all to take a share of the high-risk motorists, so that all such motorists are insured. It was started because of high costs.

There should be a similar pool of medical risks. Even if they refused actually ill patients, they could have an at-risk medical insurance pool, or life insurance pool.

6. Nursing homes are reluctant to take people with H.D. because they "take six times as much staff work as the average." There could be a tax credit or deduction for each high-cost patient (i.e., H.D.) they accept. Or, there could be a high-risk patient pool, like insurance pool, to pass out among nursing homes; but a tax cut would be more acceptable and less likely to cause cries of "government interference," and thus the H.D. patient may be viewed and treated as a more welcome guest.

Cases of all but two of enclosed people are from people who have nothing to do with the organizations fighting H.D. I wanted to tell you their stories because you need to suspect how many more people are hiding in the woods with these problems.

Of all people mentioned, two women are on mailing lists for newsletters of H.D. committees and one couple is in the membership.

Life Insurance.

We just don't mention H.D. on any insurance.

Therapy.

There's no free therapy and no programs to give therapy to people with diagnosed, permanent, "terminal" muscle deterioration around this county (Ventura).

Counseling. (In Ventura)

The only sponsored counseling is through "Social Services Family Counseling" and it is group sessions only at a graduated cost index, depending on family income. Every known H.D. family or at-risk family refuses to go to a group who commonly have never heard of it and who (the family feels) know nothing of its problems.

We only know of therapeutic success of occasional "rap" sessions in H.D. organizations and benefits of talking with other families dealing with it.

Also, only know of paid psychological counseling being used on a private basis.

Nursing Homes.

Her kids got one woman into a retirement home but once she went into the hospital (temporary) the home said they wouldn't take her back.

One nursing home said he'd taken in one H.D. patient and they "take as much care as six regular patients." He refused to consider the other person I was looking around for.

Owner of one rest home which is a retirement home, not a nursing home, told me the law makes certain requirements of a patient's capabilities before they are allowed in a retirement home instead of a nursing home:

- a. Amount of staff and nurses per capita;
- b. Patient must be able to dress herself; and
- c. If they meet these requirements and don't "disturb the other roomers too much" and have money, he'll accept them, "one or two." I have one woman in mind I'm trying to persuade to try there.

State Legislators' Efforts.

After being involved with Case No. 4, the state legislator (locally) wanted to introduce a bill to help protect H.D. patients or whatever we thought was needed if we'd just tell him.

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We told him: therapy, nursing care allowances for poor families, more meals on wheels things, counseling, research maybe tied in with state universities, publicity, and H.D. Day, and no firing people with H.D. unless really necessary.

He got to top man in California Social Welfare System. Came back completely altered. Told us, "There's programs for people like her, can only do these things on a Federal level" and "Apply to your local social worker." The woman's husband works. They don't normally qualify for Medicare unless you can prove you were in the hospital past a certain amount or spent over a certain amount of your income in the past year on medical aid. If it hadn't been for all the other illnesses complicating the woman's life, she wouldn't have qualified for medical program which now pays all H.D. medicine and all the rest of her medicines.

About three years ago, Head of California Social Welfare Program was most unsympathetic. Seemed to think all channels exist for any H.D. needs. But there's no program to assist families paying for nursing homes. That's why so many wind up in state mental hospitals--they're free.

No therapy programs are set up for "terminally ill" even though I know an H.D. patient benefitted from them when her social worker somehow pulled some strings to get her in one to see if she'd benefit.

Drugs cost money. From time to time the treatment is altered, and you buy new drugs. No plan to help families who lose a breadwinner, except Social Security, which is inadequate for a growing family.

Sterilization.

I know one woman with a hereditary skin disease, rare and uncomfortable. No one ever suggested not having children because of it.

One other woman in Santa Paula has an hereditary disease like "loose joints." Any little jar and her joints dislocate. One of her children inherited her problem and is now laid up with both legs in a cast. No one suggested that she not have any children.

The woman who has passed Niemann-Pick disease on to her daughter wasn't advised not to have children. It's true the odds against receiving Niemann-Pick disease are greater than H.D., but let's judge by principles here, not by the odds.

I feel that the reason people behave in such a medieval manner in saying "sterilize people who might litter the population with H.D." is because of the fear of anything associated with mental illness.

I feel the fallacy of saying it is for the good of the prospective parent can be explained this way.

A doctor, or a fortune-teller, tells a man he has a 50-50 chance of being hit by a truck sometime between age 35 or age 50. He is told that if he is hit by a truck at any time during that period then each of his children has the same 50-50 chance when they reach the same age bracket. He is recommended not to have any children on these grounds.

"But," the man argues, "I will be a mature man then. Many people have lived a full life by then and do die."

"But Mr. S., you may not die when the truck hits you. You may live on with physical impairments and maybe some mental ones."

"Oh, I see, it's not a life or death matter that really has you recommend this course of action. It's that you're afraid the quality of my life may deteriorate. Well, Sir, that weakens your argument considerably. Thank you. Anyway, I understand that 50% of all Americans will die of some form of heart disease, Sir, so I would strongly urge you not to let any children of yours run the risk and sterilize yourself. That is a life and death matter, especially as heart disease seems to run in your family."

Having children is not practical. It fills an emotional need. Therefore, to argue with reason and "practicality" and "odds" is unreasonable. If practical considerations were all that mattered no one in this society or economy would raise a family.

April 15, 1977

ANONYMOUS
CALIFORNIA

MARCH 27, 1977

As the husband of a victim of Huntington's disease, I wish to bring to your attention some of the problems we have faced.

About 20 years ago my wife's handwriting and walking began to deteriorate. I did not know that there was a history of this disease in her mother's family, and I don't think she did either, although we knew that her mother had died in a mental hospital. There was no evidence of mental deterioration in these early stages of my wife's symptoms, so we had no idea of the reason for the deterioration in muscle control.

For three or four years our family physician tried to cure the condition before he referred us to a neurologist. It took a year of tests, observation, and probing into family background for the neurologist to arrive at his final diagnosis of Huntington's disease. Up until that year, I had never heard of the disease. The neurologist's statements were not at all encouraging, as he told us no research in the field of this disease was being pursued. His recommendation for the eradication of the disease was that all children of victims be sterilized. Our two children were then in college, from which both subsequently graduated, one having attained a master's degree.

During the 16 years since this diagnosis, and even before, my wife's condition affected my career by causing me to avoid professional situations which might require traveling or appreciable overtime work. Another limitation was the need to stay with the employer through which we had medical insurance, as it was almost certain that no new insurance carrier would cover this existing condition.

The insurance we did have helped with the costs of physicians' services and medication. When, after bravely striving to carry on her household chores and care for herself for several years during which it became ever increasingly difficult, my wife finally required the care of a nurse during the hours when I had to be away at work, the insurance covered the bulk of the wages of the nurse.

However, when it became no longer feasible for my wife to be cared for at home, she still did not need care in the acute care section of a hospital. An extended care facility was all that was needed, but the insurance policy covered only acute care hospital costs. Finally the carrier agreed to pay their share of the hospital cost attributable to nursing services. Two years ago the lifetime limit of the insurance coverage was reached, and that financial aid ceased.

My wife did not have enough time credited to her Social Security account to be eligible for disability benefits. So far I have not found any other source of public assistance that is not predicated on a condition of virtual financial ruin. By working an average of about 65 hours per week, I have been able to stave off the condition so far, though our savings have been drastically reduced.

Financial assistance not predicated on exhaustion of savings would be of tremendous benefit to victims of all catastrophic illnesses such as this, and to their families.

My wife was a popular, musically talented girl who had graduated from high school at the top of her class scholastically and took a leading part in various activities as a young adult. For over ten years she suffered the frustrations of increasing difficulties and limitations in carrying on the activities of a wife and mother, and for the last seven years she has been confined to bed or wheelchair in an extended care hospital, being completely bedridden now. This, of course, has had a serious effect on her family, both psychologically and financially.

My wife's age is 62, mine 64, and my occupations are engineering and college teaching.

Los Angeles, California

April 15, 1977

ANONYMOUS
CALIFORNIA

MARCH 27, 1977

I am a 38-year-old married woman with an 11-year-old boy and a 16-year-old son, both at risk, as my husband, their father, has the disease.

Here is a list of problems involved with the patient and family of Huntington's disease.

Original diagnosis is often overlooked in favor of more well known nervous system diseases. In our case it involved two auto accidents before the problem was brought into focus. Next, the acceptance by family and friends both of prognosis and symptoms as well as transmission. Also the children are finding it extremely difficult now in dealing with their father's apparent lack of interest in them and mother having to take over all authoritarian roles for the family. The inability to find help in the home both from financial problems and fear of the manifestations has created enormous emotional strain as a wife, mother, or even a person. I am unable to continue my education in my field of work while I am at the same time desperately needing the extra income it would eventually bring in. I am a Medical Coding Analyst.

Financial needs for in-home health are very much in focus as are means of keeping the patient as active in the family for himself and his family.

Medicare benefits should be available sooner for any such extensive diseases.

Insurance is a very real problem for this type of patient and their children.

The children should definitely have genetic counseling without the cost being prohibitive.

The impact on a marriage is very bad as the unaffected partner is in a most trapped position for their own future, though much desiring to care for the patient. This is particularly trying for a Christian with strong views on marriage and the home, though I must admit I'm not sure I am strong enough to uphold my convictions under this constant drain on my energies, emotions and physical well-being.

Los Angeles, California

April 15, 1977

Please don't use my name publicly as it could be harmful in trying to advance my career.

April 15, 1977

ANONYMOUS
CALIFORNIA

APRIL 2, 1977

I am a close friend of _____'s wife died of Huntington's disease recently, after a 15-year siege of suffering that progressively worsened and at the end was appalling. Mrs. _____ had severe brain damage, could not talk intelligibly or control her bodily functions--but she was not a "vegetable." She flailed about helplessly, bruised and sore, and frequently screamed for long periods. But the family insisted on having her cared for at home, and surrounding her with love and warmth. When she died--of pneumonia, a complication--they sincerely mourned their loss, because they had loved her very much, and she had been a wonderful wife and mother as long as she could. All of us who are close to them had great admiration for the whole family, for their love and courage.

But the horror has not ended. Any of the four _____ children, and very likely at least two of them, may have inherited the disastrous gene. This means that what they have witnessed in their mother may happen to them...the symptoms won't reveal themselves until the children are 35 or 40 (the eldest is 32 now, I believe). Maybe they'll notice unreliable reflexes or excessive clumsiness at first. If diagnosed as Huntington's sufferers, they'll have a prospect before them of steadily deteriorating in mind and body, and of ending up as a screaming, flailing wreck of a human being.

As you know, abundant funding is made available for research into such widespread killers as cancer and heart disease. But to my mind, Huntington's disease, though somewhat rare, is far more terrible than either of these for the person affected. With the other two diseases, both pain and fear of death may be intense--but the person is still himself; can communicate his pain and fear intelligibly to others; can receive comfort and, in the end, die with dignity. The only dignity in Mrs. _____'s death was the extraordinary courage of her family and the memory of what a fine person she had been. I believe that subjectively, she experienced her last days--or should I say months--as a howling, senseless inferno of isolation.

Los Angeles, California

April 15, 1977

Please support research into a possible cure for Huntington's disease; a method of early diagnosis; drugs for the significant alleviation of symptoms.

Thank you very much.

Los Angeles, California

April 15, 1977

ANONYMOUS
CALIFORNIA

MARCH 26, 1977

Two letters recently reached my home. They were sent because my wife has Huntington's disease. The first was from the Veterans' Administration, Wadsworth Hospital Center in Los Angeles, California. There, it has the Human Neurospecimen Library (tissue bank)--the only place in the nation which stores fresh-frozen tissues for H.D. research. This organization seeks tissues from H.D. afflicted persons and from at-risk family members. It also solicits organ donations from deceased H.D. patients. It wants my wife's brain when she dies, and I will probably respond in the affirmative.

The second letter was from the Committee to Combat Huntington's Disease (CCHD); the letter said that the Commission for Control of Huntington's Disease is collecting relevant testimony on which to base recommendations to the President and to Congress. I have been asked to cooperate by sharing my experiences. I don't know if my story will do any good, but I realize that doing nothing will certainly do no good.

I have tried to describe the progress of the disease as I saw it, indicate the efforts my family made at maintaining a normal life, and discuss the problems of diagnosis, expense and treatment. It is virtually impossible to describe the family's sense of frustration and despair.

I wish I did not have the background to tell the following story, but unfortunately, I do. The story has a beginning, but at present, the ending is unknown.

BACKGROUND: In 1944, while stationed in Santa Rosa during World War II, I met my future wife, June; she was a lovely young woman of fine character. I also met her parents, her sister--younger by two years, and her brother--younger by eight years.

The war ended in 1945, and I was discharged. Immediately I enrolled as an engineering student in the University of California at Berkeley. In June 1946, June and I were married at St. Rose Church in Santa Rosa. We lived in Richmond while I attended school. Our first child, a son, was born in October 1947,

and we named him Joseph, after his maternal grandfather. At that time, I was in the junior class. June, who had been working at the U.C. Radiation Laboratory, became a full-time mother and homemaker.

A second son, Christopher, was born in December, 1948, when I was a senior in college.

At this time, June's father became ill with a low-grade fever, and was moved from a hospital in Santa Rosa to St. Luke's in San Francisco, and finally to Stanford Lane in San Francisco. Tests could not ascertain the cause of his illness, so he returned home. He was a bookkeeper but now was unable to perform his work. He had played the piano, but that, too, became impossible. A local physician diagnosed his ailment as Addison's disease, and he was treated for this for the 12 remaining years of his life.

When I graduated in 1949, we moved to Seattle where I had found employment; consequently, we had infrequent contact with June's father until we returned to California in 1957.

As I recall, June's father was placid and did a lot of sitting. Sometimes he would get angry and, on occasion, would develop a violent rage. He lived at home. His wife took care of him, and his son provided for them. In 1960, after a short stay in a hospital, he died of a cerebral hemorrhage--probably caused by hitting his head in a fall. The hospital physician thought he might have had schizophrenia rather than Addison's disease, but later we came to believe he had Huntington's disease.

While in Seattle, we were blessed with two more children: Timothy in 1952, and Susan in 1956. We now had four fine children.

In 1957, we moved to Sunnyvale, California. June was 34 years old. She had varicose veins since the birth of our first child, and sometimes had lower back pains; otherwise, she was in good condition.

It was around 1954 when June evidenced fear or riding in the car on mountainous roads--particularly going downhill. This fear slowly became more pronounced with time.

June remained busy with the children. She carpooled with other ladies in taking children to catechism classes. In 1965,

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While taking Susan to her piano lesson, June stopped, parked the car, walked home, and never drove again. Later, I believed she no longer could make decisions about driving the car. She was 42 years old at that time.

At this time, Joe graduated from high school and started college. Chris had ulcerated colitis five years earlier and had another serious attack the previous year. Both times, he was in the hospital for extended periods. We were very concerned.

About 1968 or 1969, June apparently was having her menopause; she was 45. The doctor prescribed Premarin.

In 1969, June became inactive, sitting quietly in preoccupation. She also began to lose weight and didn't look like her natural self. I thought it might be the change of life, and I suggested she see a doctor. She refused. June had always tried to avoid seeing doctors and dentists.

Joe graduated from college in 1969, received his ROTC commission, and started a semester of postgraduate work.

Until now, we thought we had been an average American family with success, disappointment, health, illness, and emphasis on the importance of family life. We had a fine 1969 Christmas with the family together, but June was ill.

In 1970, problems began to increase.

In January 1970, we persuaded June to have a checkup with the doctor. Kenneth, the doctor, was a general practitioner. He noticed that June had deteriorated in mind and body, and he placed her in the hospital for various tests and for examination by a radiologist and by a neurologist whom we will call Jerrod.

After six days in the hospital and several examinations, we were told that no physiological problems could be found, and psychological examination was suggested. We spent a month adjusting to this; March arrived.

Meanwhile, I was starting a new project at work, and I became increasingly busy. Work forced me to travel occasionally. June didn't like this. In early 1970, I was traveling to New Mexico, and June became very nervous.

Chris was living at home. June asked him to drive her to Santa Rosa to visit her mother. She rode in front with Chris, but after 40 miles of driving, she suddenly threw herself into the back of the car and became extremely upset. Chris found a hospital in South San Francisco where they treated her with Thorazine, and subsequently Chris brought her home. He called me in New Mexico. I immediately flew home. June was almost 47 years old. The children were 22, 21, 17, and 13.

June was still in a daze from Thorazine when I got home. I phoned the doctor who arranged for further hospital examinations. Again, no physiological causes were found, and psychiatric care was recommended. We were frightened.

After June came home, we contacted a psychiatrist who suggested further examination in the psychiatric ward of the hospital.

Here was a new and alarming experience; we were apprehensive--especially June; she didn't want to go to a strange place with a lot of "crazy people."

We were fortunate; a priest, Father Al, visited us and discussed our problem. A man of compassion, Father Al had been a seminarian and taught religion when Chris was first ill. He offered us strength then, and now he reassured us that the psychiatric ward was not unpleasant, and that we had nothing to fear there.

In April, June spent 12 days in the ward for tests and examinations. We never learned what was happening with June or what was ascertained.

After June came home, we saw the psychiatrist regularly--once or twice a week. He asked questions; we talked; he listened; we learned nothing! After two months, he recommended a different psychiatrist who used another method of treatment. He also told us that psychotherapy had been ineffective so far, and that June should return to the hospital for possible diagnosis of a slow-growing tumor, genetic brain damage, endocrine gland malfunction, and for further psychiatric analysis. He suggested that there were too many signs of psychiatric disturbance for the causes to be clinical or biological, and he further suggested we consider shock treatments or prefrontal lobotomy--depending on consultation.

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The psychiatrist was concerned about how the family might react to the situation if there were no improvement, or if June's condition worsened. Suggested options included:

1. Home care, depending on June's condition and our ability to take care of her;
2. Hospital care and therapy, which would cause financial ruin;
3. State hospital care.

The situation was almost of crisis proportions, and none of the options were attractive.

At the end of May, June returned to the hospital psychiatric ward for two months. In some ways, she liked it there. We also engaged another psychiatrist on the recommendation of the first one. The new psychiatrist was named Allen; he was assisted by Pat, a lady psychologist who practiced Gestalt psychology. Hospital examinations seemed to substantiate the belief that June had a psychiatric problem.

June came home near the end of July. We both began regular visits to Pat once or twice a week; later, Allen would join us. June seemed to be helped by this therapy, and she liked Pat. Allen eventually became my friend, confidant, and advisor. He provided the continuity needed in medical counseling.

Joe went on active duty that year and was assigned to duty in Germany. He left in the fall. We regretted seeing him go, but we were glad he wasn't sent to Viet Nam.

Chris finished junior college and went to the University of California in Berkeley. We missed him, too, but he came home regularly.

Tim finished high school and began junior college. Susan entered high school. Susan was a great help to us, but I had to be careful that she didn't become the "housewife" and miss being a young girl.

Things went reasonably well for about four months with therapy sessions and June taking Valium. We had a few tense sessions at home. I tried to avoid trips, but when they were absolutely necessary, Tim and Sue looked after things while I was gone.

June had increasing difficulty in making decisions; even selecting ingredients for a sandwich became a task. Her condition worsened.

The financial problems for this situation can be enormous, but fortunately my employment benefits included a medical plan with psychiatric coverage. Medical costs for the year were \$10,300. Insurance paid \$8,000, and we paid the remaining \$2,300. The latter was a heavy financial burden, but we managed.

THE GENETIC LEGACY

In December, June's upsets became much worse and they were contagious; we were all on edge. In January 1971, it was necessary for June to be in the hospital for 28 days, and for another seven days in February.

Jerrod suggested that while June was in the hospital, he take a pneumoencephalogram. It was almost an afterthought, and he doubted much would be learned. The encephalograph showed extensive atrophy in June's brain. Jerrod said it might be Huntington's disease, and that was the first time we encountered this term.

Jerrod and Allen explained Huntington's disease to me. I neither understood nor accepted it at that time. I remember phrases like "progressive," "no cure," and "hereditary." June was angry at Jerrod for telling her something she did not want to hear. Chris, who was majoring in biology, explained to me that when the affected cells die, they can't be replaced. Even before I could absorb the impact of the tragic news, I began searching for something different than the psychiatric ward. In Santa Clara County, where we lived, we found nothing, but in the next county we found a rehabilitation center which seemed to offer a good program. Because it was out of our own county, the services could not be provided free to us; insurance did not cover it; this meant we would pay the bill. We decided to try it; why, I'm not sure.

At the center, June partook of exercise and craft therapy. Visits were continued to Allen and Pat. At the center, June was heavily doped with Valium, but she seemed to enjoy her stay there. However, the cost was ruinous, so we reluctantly brought June home.

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June's mother, sister, and brother, as well as we, were anxious to determine whether or not it was actually H.D. Jerrod attempted to trace the illness of June's father. He checked relatives' recollections of family illness. The post mortem on June's father was useless, and relatives denied knowledge of such illness in the family. The parents of June's father came from Italy, and it was impossible to trace further.

The remainder of the year, June stayed at home and there was a minimum of upsets. We accepted the problem and even made a few short trips for visiting. June rode in the back seat and still experienced anxiety when we drove on hills.

June's mother and my mother came for visits, but the visits usually ended with more upsets. June was inclined to insist that she was doing fine and even improving. She was determined to be well. She was 48 years old.

We continued therapy with Allen and Pat, but sometime during this period, Allen said that he was more concerned about me than June. This, I could not comprehend; I considered June's welfare as our prime objective.

Joe sent June a bottle of water from Lourdes, and she put drops on her head until the water was gone. She also wore out two sets of rosaries. Father Al had been transferred, and his replacement nor the Pastor didn't find time to visit June. This made me angry!

Joe came home just after Christmas in 1971, and we were glad to see him. Joe had been the only member of our family unable to be with us on our 25th wedding anniversary that year.

I can recall that in the late 1960s, June had facial twitches. As her recognized illness progressed, additional problems of coordination and involuntary muscular movements became worse. It also was more pronounced when she got excited. In early stages of her illness, it was not a problem, and we traveled as we wished.

During 1971, medical expenses were about \$7000; I paid \$3,400--an increase from the previous year.

In January of 1972, periods of upset and depression caused such family irritation that June went to the hospital for a month

and a half. The Christmas Season seemed to make her worse. The hospital resisted accepting June. Her illness, they stated, was not the type they treated, but Allen justified it on the basis of the well-being of the entire family.

Allen suggested we place June in a convalescent hospital, but I couldn't agree to that. He also suggested we make out wills, have June declared incompetent, and put her into conservatorship. This was logical because June often became irrational and even threatened divorce. To avoid further complications, we had an attorney prepare our wills and the conservatorship, painful though it was.

Things continued in a turbulent state to which we had become conditioned.

Chris graduated from college and was vainly looking for work. Tim went to college in Berkeley, and we missed him. I was given a new assignment with more responsibilities at work.

In the fall of 1972, I realized that the continued state of stress and the extreme demands of the situation could not continue. I reluctantly began to look for a suitable convalescent hospital.

Convalescent hospitals are a jungle to the novice. There is residential care, intermediate care, and full care. I decided intermediate care was most appropriate because it included therapy and exercises.

Most convalescent hospital populations are old, incapacitated, and passive--very depressing. At that time, prices varied from \$15 to \$20 per day.

I decided the best place was not a convalescent home, but a residential care home I had found run by Catholic sisters. People were moving about and didn't seem sick. Still, I couldn't make the move.

June got worse and went back to the hospital for a few days. After that we tried the residential care home, but the other residents demanded June leave because her conduct was abnormal.

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Shocked and disappointed, I brought June home. She was both hurt and pleased.

A month later, we again placed June in the hospital, but they insisted she couldn't continue to come there. The hospital staff, whom June really liked, marched her to an interim convalescent home amidst June's protestations.

After a month in the convalescent hospital, June was moved to another place more suitable and financially viable. June had become accustomed to the convalescent hospital, and didn't seem to mind being there. We could visit her frequently and bring her home for weekends. June soon made friends with two ladies at the hospital, adjusted very well, and again displayed a different personality than she did at home. This condition continued into 1973. We brought her home on weekends, but not as frequently as previously.

A few years earlier, June had used Haldol medication, but it seemed to make the symptoms worse. In 1973, Jerrod told us that in England persons with H.D. had taken lithium carbonate which had reduced movements, so we decided to try it. I don't know whether it affected the H.D. or just alleviated the depressions, but June seemed to be doing better. In October 1973, we brought her home; it was a happy occasion for all of us.

The remainder of 1973 and 1974 passed comparatively smoothly. We managed to go to a few places and enjoy our family. Sue graduated from high school in 1974 and got a job. She and her boyfriend planned marriage. Things at work started to go sour for me. I didn't know whether or not it was my fault, but I felt insecure. Tim had moved back home and was commuting to Berkeley. Chris still hadn't found a decent job, so at his invitation he and I went into business together; this was my backup employment and his primary job.

In January 1975, Sue was married. Tim finished college and went into business with Chris--we planned to expand.

June and I were alone now. I decided she needed more attention, so I employed a woman to stay with her four hours a day during the week.

In May 1975, Chris was married--another happy occasion. June had trouble walking down the aisle and remaining still for photographs, but she succeeded.

June's difficulty in walking and the involuntary movements were both increasing. It was just after Christmas 1975 that June lost her balance and toppled over backward striking her head. It was obviously a hard blow.

Chris' wife was four months pregnant. June and I both wanted grandchildren. I have never known anyone to love babies as much as June. I prayed that she would see her grandchild. It is a moot question as to why the children would risk having babies knowing the possible H.D. risk. Fully aware of the terrible potential as evidenced by June's experience, they made their own decision.

I had changed parishes, and now Father was visiting June every month. It made us both feel better.

Tim was married in March 1976--another happy occasion. And Joe went into business in another city.

Our grandson was born in May; this brought new light into our lives.

June fell more frequently now, and I was afraid to leave her alone at all. I came home from work one evening and found her washing blood from her head after a severe fall. I decided it wisest to put her back into a convalescent hospital.

Allen arranged a hospital checkup for June before she went into the convalescent hospital. June didn't want to go, and I cancelled the plans twice before I finally had the courage to again make the move. It was difficult to make that decision. I knew she would never live at home again.

Allen and Jerrod decided lithium wasn't doing any good and discontinued it, but continued the Valium.

Shortly after June went into the convalescent hospital, I saw in the morning newspaper that Allen had died. I was shocked. We had lost a friend upon whom we could depend, and with him had gone my continuity in the medical world.

June continued to fall in the hospital. The staff was concerned, but she insisted on moving around. She also became incontinent, and that embarrassed her. We visited her regularly and brought her home for weekend visits. She continued to fall at home, but insisted she was alright.

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June has a healthy appetite and loves milkshakes, so I take her one almost every evening. Sue visits her almost every day. Joe is here for a visit every week, and Chris and Tim visit when they are in town. June loves to see the children and the baby, but she usually cries when they visit her. The hospital staff is very good to June.

In a recent fall, June hurt her leg and broke her collar bone. Now she is in light restraint and stays in bed. She can hardly walk. Fortunately, she is quiet while lying down. We brought her home in February for my birthday. Her mother and sister were here. June was so happy and got so excited that she wouldn't stay in bed and couldn't relax; Joe and I had to take her back to the hospital. It may have been her last visit home.

Both Sue and Tim's wife are expecting babies in July. June will be 54 in June. I pray she will be able to enjoy these youngsters.

This is a story that goes on and on with new episodes and an indeterminate ending. The prospect is unattractive, but we will continue to do the best we can. We have many blessings to help us endure the adversities; with God's help, we shall continue to do so.

A BACKWARD LOOK FOR ENLIGHTENMENT

I have a vested interest in H.D., its diagnosis, treatment and cure. My family and my wife's family also have vested interests in the very same things. The amount of interest is illustrated, though inadequately, by the figure which is part of a family tree. Two H.D. patients are shown along with six persons at risk and a number (which hasn't stopped growing) of others whose risk is determined by status of at-risk persons; i.e., who has the bad genes?

1. Rare--The Disease or the Diagnosis?

My experience indicates that diagnosis is difficult and perhaps rare, even though we are surrounded by medical excellence.

- a. Doctors need to be more aware of H.D., its symptoms and methods of diagnosis.

- b. Research must develop a test which will detect H.D. victims and latent H.D. victims, perhaps down to the fetus level.

2. Ignorance of the Medical Maze

I found that I was ignorant in a number of areas and that, for one reason or another, it is difficult to get answers. Medical questions for the layman include What is happening? What is likely to happen in the future? And what can be done?

The latter question leads to where do you find the answers, the experts, and the facilities? I don't mean to imply that a person needs a medical wet-nurse, some things one must do for themselves, but there should be an available source of such information as it is needed.

- a. A simple medical guidebook is needed to describe what H.D. is all about, the possible or typical course of the disease, and what kinds of treatment can be considered.
- b. At the local level, a guidebook or "yellow pages" is needed to locate experts in the field, if any, and facilities such as hospitals, home nursing services, etc.

3. Research

The purpose of research is to shed light on the unknown. Achievements can't be scheduled nor ensured. It should be funded steadily and at a reasonable level.

- a. Basic research is needed to understand the origin and vehicle for H.D. and from this tests and cures will be possible. A dedicated program in H.D. research is required which includes interchange with research in similar areas.
- b. A program of investigation of potential treatments for H.D., its symptoms, and its progress is needed to improve the lot of those suffering from it.

April 15, 1977

4. Treatment

I am not aware of any effective treatment for H.D. although various drugs have been tried with various claims of success. At the present time, it seems that patience, kindness, and love are most important along with some program of activity.

a. Recommendations--you tell me!

5. The Mind and The Soul

As you recall, we got started off with psychiatry before H.D. was diagnosed, and continued psychiatry after the diagnosis. I believe it was a considerable aid. H.D. is a disturbing mental factor both to the afflicted and the family.

a. Psychiatrists and psychologists should be aware of H.D. and its impact on the afflicted and the affected.

b. Faith in God is a source of inner strength, a resource which has at least benefits equal to psychiatry for those fortunate enough to have a personal belief.

6. Financial

I was fortunate enough to have a good medical insurance program, the additional ability and desire to take up the slack, and the sense not to attempt what we could not afford if it offered no great promise.

Often, the ground rules of insurance or government-supported programs dictate the course of treatment to the detriment of the patient and at increased cost. For example, if only hospitalization is covered, the patient can go to a hospital at high cost but can't go to a convalescent hospital at lower cost or have home care at perhaps even lower cost--also a more desirable situation.

a. Medical insurance carriers should review their policies to make them more flexible, more efficient and more effective.

- b. Aid should be given to people to help them continue a productive life rather than encouraging them to exhaust their savings and accept welfare.

April 15, 1977

ANONYMOUS

GARDENA, CALIFORNIA

MARCH 10, 1977

Many times I've tried to write this letter. I didn't want to remember the hell of the past twelve years; I don't want to think of the hell to come if nothing is done, if a cure is not found.

My husband has Huntington's disease. I have six children who are at risk, and I have three grandchildren. My children's ages are 21 to 31. TIME IS RUNNING OUT!

We had a good life. My husband was an attorney. We were a close knit family. I watched my world fall apart little by little. There were flairs of temper, unreasonable demands, calls from his law partner, from clients, from collection agencies, from the Internal Revenue.

I do not have the words to describe the next five years. We started our rounds of doctors only to be told there was nothing wrong. After being told this by several doctors, I reasoned that if nothing was wrong with him, then it must be me. I tried to commit suicide. I became the patient under psychiatric care. Finally, we got the diagnosis of Huntington's disease after it became obvious that something was wrong with my husband.

H.D. had not only ruined us financially, but it caused financial loss for many of his clients. Worst of all, H.D. destroyed our family emotionally. Two of our children had nervous breakdowns. We all had to undergo psychiatric help individually and as a family.

It's twelve years later, and the nightmare goes on. My husband has been in a state hospital for the past seven years. Five out of six of my children say they will commit suicide before they go through the agony they have seen their father go through or be the cause of others that love them to have to go through.

Aside from the catastrophic impact Huntington's disease has on H.D. families, there is the practical side. If you could make our legislators aware of the amount of money it

takes to care for the victims, and their families, they would realize the practical benefit of starting an intensive research program. It would not only give those affected by H.D. hope, it would save the taxpayers millions of dollars.

This is a letter my eldest daughter wrote several years ago:

"I am 27 years old and have three children. My father has Huntington's disease.

"Daddy was diagnosed three and one-half years ago and for a long time I could barely talk about it. All the fears, the panic had to stay inside because I didn't want anybody to know how scared I was.

"I watched myself constantly for signs and symptoms of H.D. and I was always finding them--in any awkward movements, any memory lapse, any twitching muscle, any outburst of temper, any anything. I was sure I had the disease. But I couldn't talk about it. I didn't want anybody watching me for symptoms as I watched myself. I didn't want anybody feeling sorry for me. I didn't want to go to a mental hospital.

"The fear of H.D. wasn't the only thing tormenting me. I had to deal with my feelings about my father and the way he was forced to live in a horrible institution that was more like a prison than a hospital. While the disease was slowly attacking his brain and body, the hospital was immediately crushing his spirit and robbing him of any hope.

"I've always loved my father. He was so special to me and I was special to him. But I couldn't do anything for him--except visit. I felt that as I watched him deteriorate, I watched myself. I wanted to take him home with me and take care of him. But I had three children under four years old (the youngest with brain damage from birth) and no money and was separating from my husband. I knew I couldn't take care of him. I wasn't real sure I could take care of myself.

"Of course, I wasn't the only one involved in this ordeal with my father. My mother and five sisters and brothers were in the same nightmare. But we couldn't help each other and, in fact, we nearly tore each other apart with our fears, guilts, and demands.

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"I attacked my mother for abandoning my father in the hospital, for not taking care of him. In my own pain and helplessness, I couldn't see the hell my mother was going through. I resented my brothers and sisters for not visiting Daddy enough. They resented me for the pressure of demands I made and for my unyielding insensitivity to them."

April 15, 1977

ANONYMOUS
CALIFORNIA

APRIL 27, 1977

I am a 34-year-old woman, at risk, single, but living with a man for the past eight years. For the past five years, I have been a professor of history at a state college in California. My mother was diagnosed with H.D. eight years ago. There is one point I would like to make to the Commission: families who discover that one member has H.D. should allow those who are at risk to feel miserable, depressed, crazy, wild, whatever they like, and to know that it is all right to feel that way. In my own experience, after learning that my mother had the illness, I tried to go right ahead with my life (at that time I was taking Ph.D. exams and beginning to write a dissertation) as if nothing had changed, as if nothing were different.

I had a great deal of trouble with my dissertation, and with the man in my life as well, but I was never able to connect all of that with my mother's illness. All I knew was that I was not happy, that my research was going badly and the writing of my thesis worse. This surprised me, since I had done very well in graduate school up until that time. I wish now that someone had been able to point out to me the possible connections between the difficulties I was experiencing and the illness, and that I had been able to feel that I had a right to feel miserable, that my depression and confusion were legitimate feelings.

I had many dreams at the time (which I wrote down) about death, illness, being pursued by murderers, but again was totally unable to make any connection with the illness. Only now, eight years after my mother's diagnosis, am I able to understand the feelings that I had then. I would hope that those who are in a position to counsel at-risk individuals, and the parents of at-risk people, would communicate to them that it is all right to feel very miserable and sad and depressed when confronted by a tragic illness.

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Reports of the Commission for the Control of Huntington's Disease and Its Consequences

Volume I: Overview	<i>DHEW Pub. No. (NIH) 78-1501</i>
Volume II: Technical Report	<i>DHEW Pub. No. (NIH) 78-1502</i>
Volume III, Part 1: Work Group Reports — Research	<i>DHEW Pub. No. (NIH) 78-1503</i>
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Information on how to obtain additional copies of the Commission's reports is available from:

National Institute of Neurological and Communicative Disorders and Stroke
Office of Scientific and Health Reports
Building 31, Room 8A08
Bethesda, MD 20014
(301) 496-5751



Information about Huntington's disease is also available from the NINCDS office as well as from the following voluntary health organizations:

<http://nihlibrary.nih.gov>

Huntington's Disease

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9701 Wilshire Blvd.
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